

What do People with Down Syndrome want from their Cardiac Team?

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Table of Contents

Acknowledgements.....	ii
Table of Contents.....	iii
List of Tables.....	vi
 Abstract.....	 1
 Chapter 1 – Introduction.....	 2
An Introduction to Down Syndrome.....	4
Congenital Heart Conditions.....	8
Health Provision for People with Down Syndrome.....	10
Health Research and Down Syndrome.....	12
Why This is an Important Area to Explore.....	14
Barriers to Research in the Area.....	15
Summary.....	17
 Chapter 2 – Methodology and Method.....	 19
Participants.....	20
Recruitment.....	22
Rationale for Qualitative research	22
Thematic Analysis.....	23
Theme Development.....	23
Stages of Thematic Analysis.....	24
Phase 1 – Familiarisation with the data.....	24
Phase 2 – Generating initial codes.....	25
Phase 3 – Searching for themes.....	25
Phase 4 – Reviewing themes.....	26
Phase 5 – Defining and naming themes.....	26
Phase 6 – Producing the report.....	26
Ethics	27
Ethical Approval	27
Consent	27

Ethical Risk.....	27
Confidentiality.....	28
Reflexivity	28
Advantages and Disadvantages of Research with People with a Learning Disability.....	30
Interview Design.....	31
Epistemology.....	33
Chapter 3 – Findings and Discussion	35
Theme 1. Who is the Patient.....	37
Theme 2. Self-Care, Care from Others and Gaps in Care.....	47
(i) ‘I Do it Myself’ – measures taken to manage health.....	47
(ii) Help from others – the regular input of others to manage health difficulties.....	53
(iii) Emergency admissions and gaps in care.....	56
Theme 3. Health and Fitness.....	59
(i) ‘Breathless and Dog-less’ – implications of heart and health on fitness.....	59
(ii) ‘Its Part of a Strict Diet’ – eating for health.....	62
Chapter 4. Implications and Limitations.....	66
Implications.....	67
Limitations.....	70
Chapter 5. Critical Reflections.....	75
“Don’t have a Scooby” – making consent meaningful.....	76
References.....	83
Appendices	
Appendix A.....	99
Appendix B.....	102

Appendix C.....	104
Appendix D.....	106
Appendix E.....	108
Appendix F.....	111
Appendix G.....	113
Appendix H.....	114
Appendix I.....	118

List of Tables

Table 2.1.....	21
Table 3.1.....	36

Abstract

This research investigates the experiences of people with Down syndrome who have a congenital heart condition (CHC). While people with Down syndrome are at greater risk of CHC, little is known about their views on hospital treatment and healthcare. A review of the literature indicates the incidence and implications of CHCs for people with Down syndrome including barriers accessing health care and higher mortality for this group. The research considered ethical issues pertinent to conducting research with people with a learning disability including the choice of research method, participant recruitment, data collection and analysis. A qualitative approach was adopted and involved semi-structured interviews with five individuals with Down syndrome and a CHC. The interview covered topics including in the impact of Down syndrome, the impact of a CHC, and involvement in healthcare decisions. Three themes resulted from a thematic analysis: Who is the Patient; Self-Care, Care from Others and Gaps in Care and Health and Fitness. The major implications of this research for people with Down syndrome, their families and healthcare professionals include the need for clarity regarding supported decision making and supporting those with Down syndrome who may not have family members to aid decisions. This research involved a small relatively homogenous group of individuals and the area could benefit from further research, for example those without parent carers to support healthcare decisions and medical procedures. The research highlighted difficulties inherent to carrying out research with vulnerable groups, such as those with a learning disability. Future research should consider the process of including vulnerable groups in research, such as how to balance the research agendas of the researchers and the researched.

Chapter 1. Introduction to Research in the Area of Down Syndrome and Congenital Heart Conditions

The area of health and learning disability has seen significant changes in recent years. Historically, there was a view that disability and illness were one and the same; people with a learning disability were presumed to have poorer health than the general population and health intervention focused on bridging the gap caused by the disability (Krahn, Hammond, & A. Turner, 2006). Such views have changed, however, with social models of disability contributing to a better understanding of the impact of the environment on health, disability, and the individual (Drum, Krahn, Culley, & Hammond, 2005). The twentieth century saw the recognition of the right of all citizens to good health, defined as a state of complete physical, mental, social well being, and not merely the absence of disease (World Health Organization, 1946). People with a learning disability have been moving towards this ideal through social movements including normalisation (Brown & Smith, 1992), and educational inclusion (Ainscow & Booth, 1998), ensuring that people with a learning disability become more visible and are integrated meaningfully into their communities.

Definitions of disability have subsequently changed, moving away from definitions that labelled the whole individual as 'handicapped' or 'mentally retarded' and towards an understanding that disability is only one aspect of a person. Definitions now reflect that the health and well-being of each individual is influenced by a wide range of factors as disability is "...complex and multi faceted, with its roots in culture" (Mont, 2007, p.1662). A change of definitions does not, however, necessarily remove the negative connotations that labels can bring to bear (Hastings & Remington, 1993). Mencap, the largest learning disability support organisation in the UK, adopt a broad definition to define learning disability:

People with a learning disability find it harder than others to learn, understand and communicate. People with profound and multiple learning disabilities (PMLD) need full-time help with every aspect of their lives - including eating, drinking, washing, dressing and toileting (Mencap, n d, What is a learning disability?, para. 4).

In order to access learning disability services in the UK, however, individuals need to show demonstrable impairment in intellectual functioning; difficulties in social behaviour, adaptive skills, or communication; and an onset before the age of 18 (British Psychological Society [BPS], 2000). Also termed intellectual disability, it is

classified by the American Psychological Association (American Psychological Society [APA], 1994), according to four degrees of severity: mild; moderate; severe; and profound, and can be diagnosed in addition to the presence of another disorder such as Down syndrome. In the UK diagnosis will generally be carried out by an educational or clinical psychologist. Clinical psychologists from National Health Service (NHS) community learning disability teams will also be involved in supporting people with a learning disability and their families to overcome the impact of the disability, helping them better access their community. Clinical psychologists may also provide support to people with a learning disability who are experiencing mental health difficulties (Deb, Thomas, & Bright, 2001). Mental health difficulties in people with a learning disability have been associated with the experience of multiple life events including illness and injury (Owen et al., 2004), therefore, it is important to minimise exposure to potentially traumatic life events and illness in this population.

Disability services in the UK are working to minimise trauma and promote independence and greater responsibility, through, for example, deinstitutionalisation and personal budgets. The challenge for health services will be to promote the health and well-being of people with a learning disability and chart the effectiveness of such promotion in terms of improvements to health, well-being, and quality of life (Noonan-Walsh & McConkey, 2009). In particular, it will be necessary to ascertain whether in addition to minimising the impact of a disability and reducing mental health risks, measures taken to improve health and well-being help individuals overcome the social barriers disability brings (Emerson, 2007; Emerson & Hatton, 2007), and the disparity of access to healthcare experienced by this group (Cumella & Martin, 2004; Jansen, Krol, Groothoff, & Post, 2006). Information as to gains made and whether these gains are significant will rely on research into outcomes at both the population and individual level, which includes those central to the process – people with a learning disability.

An Introduction to Down Syndrome

Down syndrome is the most commonly identified genetic cause of learning disability and is diagnosed by chromosomal analysis. Analysis occurs prenatally if there are risk factors present and parents consent to an amniocentesis, or antenatally following the identification of physical characteristics in babies that indicate

the potential presence of the condition. The high level of prenatal detection and subsequent termination of pregnancies make it difficult to estimate the frequency of Down syndrome (Roizen & Patterson, 2003). Evidence from various countries indicates that as the mean age of pregnancy has increased over the last 20 years, so too has the number of fetuses with Down syndrome. Research from Germany, Israel, Singapore, Australia, and the United States of America report that the number of terminated pregnancies has increased and the prevalence of live births of babies with Down syndrome has decreased from 1 in 1000 to 1 in 700 in twenty years (Roizen & Patterson, 2003).

The most recent statistics from the United Kingdom show that the number of fetuses diagnosed with Down syndrome increased from 1075 in the year April 1989/March 1990 to 1843 in the year 2007/2008 (Morris & Alberman, 2009). Of those diagnosed in 2007/2008, only 4.8% resulted in live births (91.5 % termination, 2.7% miscarriage and still birth, 1% unknown outcomes), resulting in a small increase from 4.3% live births in 1988/1989. Morris and Alberman (2009) estimate that the increase in mean age of mothers should have resulted in an increase of 48% of live births of children with Down syndrome. Screening, however, has resulted in a decrease in live births of 1% over the last 20 years (755 to 743 live births). This equates to 1.08 per 1000 births (Morris & Alberman, 2009). Characteristics seen at birth that are indicative of Down syndrome include; brachydactyly (short finger length); broad hands; brachycephaly (flat head); flat nasal bridge; hypotonia; wide first to second toe gap; open mouth; epicanthial folds; duodenal atresia; and fifth finger clinodactyly (curvature) (Roizen & Patterson, 2003). A learning disability is also characteristic of Down syndrome and children with Down syndrome will generally meet developmental milestones behind peers without the syndrome.

In 95% of cases of Down syndrome is caused by a non-genetically inherited full chromosome 21 trisomy (three copies of the 21st chromosome instead of two) (American Academy of Pediatrics [AAP], 2001). In a further 3-4% of cases it is due to an unbalanced translocation. Translocation happens when breaks occur in chromosome 21 and another chromosome, usually chromosome 14. The subsequent rearrangement of material results in some chromosomally normal sets and some with extra 21st chromosome material. Approximately three quarters of these unbalanced translocations are de novo, and one quarter are hereditary. The

final 1-2% of cases of Down syndrome are due to mosaicism, where two cell lines are present - a mixture of normal sets of chromosomes and chromosomes with trisomy 21. Individuals with mosaicism may be affected less severely than other forms of the condition (AAP, 2001; Juan, Pereira, & Souza, 2000)

Along with the physical characteristics that accompany the diagnosis of Down syndrome, comes an increased risk of CHCs (50%); hearing loss (75%); middle ear infection (70-50%); eye disease (60%) including cataracts (15%) and severe refractive errors (50%); obstructive sleep apnoea (75-50%); thyroid disease (15%); gastrointestinal atresia (12%); acquired hip dislocation (6%); leukaemia (<1%); and Hirschprung disease (a medical condition where nerve cells at the end of the bowel are missing and surgery is required to alleviate chronic constipation, <1%), (Roizen & Patterson, 2003). From an early stage particular attention needs to be paid to sensory difficulties and hearing loss and ophthalmic disorders are thought to increase with age. In addition to their genetic predisposition to physical ill-health, a sedate life style and poor diet is likely to contribute to the reported higher levels of arthritis; obesity; diabetes mellitus; and seizures in people with Down syndrome (Braunschweig et al., 2004; Roizen & Patterson, 2003). However, the relationship between this sedentary lifestyle and health is not a simple one, Frey and colleagues (2008) suggest instead that parental overprotection is the reason for lower levels of physical activity seen in youths with intellectual disability compared to their siblings. Also, a research review has indicated that children and adolescents with Down syndrome were less likely to engage in physical exercise programs (González-Agüero et al., 2010).

There is a body of literature documenting the possibility of avoiding ill-health through health promotion and training of care providers (Melville et al., 2006), increasing activity levels (Barnhart & Connolly, 2007), and working on obesity prevention. Roizen and Patterson (2003) outline how a lifelong regime is required to beat obesity and how in adults lower body-mass-index correlates with access to social activities, leisure activities, and satisfaction with friendships (Fujiura, Fitzsimons, Marks, & Chicoine, 1997). However, the wider implications of improved physical health should also be examined. The increased life expectancy of people with Down syndrome documented over recent decades (Janicki, Dalton, Henderson, & Davidson, 1999), most often attributed to improved access to medical care and

surgical procedure, however, other potentially implicated factors include deinstitutionalisation and living conditions for people with a Down syndrome and their families across countries (Noonan-Walsh & McConkey, 2009). A greater understanding of the complex medical and psychological risks that people with Down syndrome face is being achieved not only through modern medicine, but through careful research with individuals and their families.

Recent advances to the research base in Down syndrome have included an improved awareness of the complex relationships between life events, physical health, (e.g. urinary incontinence), and the presence of mental health difficulties (DesNoyers-Hurley, 1998; Mantry et al., 2008; Scior & Grierson, 2004). There is also evidence pointing to the co-morbidity of autistic spectrum disorder in individuals with Down syndrome (Howlin, Wing, & Gould, 1995), however diagnostic overshadowing can occur and symptoms of autism reported by parents are often ignored resulting in a delay to intervention and increased social and behavioural difficulties for individuals (Rasmussen, Börjesson, Wentz, & Gillberg, 2001). A lack of research in the area of dual Down syndrome and autistic spectrum disorders, particularly in adolescents has been noted (Dykens, 2007). Undiagnosed difficulties in people with Down syndrome will have an impact on the ability to appropriately identify medical symptoms and access appropriate medical care (Howlin et al., 1995).

There is also significant literature pointing to higher incidence of Alzheimer's dementia. The reported rate of lifetime incidence of dementia in Down syndrome is estimated to be between 17-21% (Coppus et al., 2006; Margallo-Lana et al., 2007), and although it is agreed that incidence increases with age, variable rates have been reported. Margallo-Lana and colleagues report clinical evidence of dementia in 50% of people with Down syndrome over the age of 60 in their UK sample (2007). Coppus and colleagues (2006) report a decrease in prevalence rates of dementia from 32% in the 55 to 59 age bracket to 25.6% in those over the age of 60. However, statistics from the United States suggest that signs and symptoms of dementia are seen in 75% of people with Down syndrome over the age of 60 (Roizen & Patterson, 2003). Signs of dementia frequently seen include seizures (58%); changes in personality (46%), focal neurological signs (40%), apathy (36%), and loss of conversational skills (36%) (Roizen & Patterson, 2003). There is also evidence of a link between early menopause and dementia in women with Down syndrome

(Cosgrave, Tyrrell, McCarron, Gill, & Lawlor, 1999). Early identification and diagnosis of difficulties will be key to ensuring access to treatment and carer support. The identification of these symptoms and noticing changes in people with Down syndrome relies on good communication and well informed paid and unpaid carers, and primary and secondary care providers who are aware of the increased risks and the latest available treatments.

Congenital Heart Conditions

Congenital Heart Conditions (CHC) are seen in 39-50% of individuals born with Down syndrome (Frid, Anneren, Rasmussen, Sundelin, & Drott, 2002; Seale & Shinebourne, 2004; Vis et al., 2009). CHCs are more frequently seen in female compared to male, and Black compared to White, babies diagnosed with Down syndrome (Freeman et al., 2008). Mortality is higher in people with Down syndrome and a CHC compared to those without (Frid, Drott, Lundell, Rasmussen, & Annere, 1999; Hayes et al., 1997; Leonard, Bower, Petterson, & Leonard, 2000; Yang, Rasmussen, & Friedman, 2002). Children with Down syndrome and a CHC under the age of 10 years have twice as many hospital admissions as children with Down syndrome who do not have such conditions and the impact of surgery will follow people into adulthood (Frid et al., 2002). CHCs most frequently seen in people with Down syndrome relate to particular physical malformations of heart including:

- Atrioventricular septal defect (AVSD, 45%): holes between both the upper two and bottom two chambers of the heart result in increased pressure within the heart (pulmonary hypertension) and increased blood flow to the lungs.
- Ventricular septal defect (VSD, 35%): a hole between the bottom chambers of the heart allows oxygenated blood to flow across the chambers and back to the lungs in addition to the normal flow. It also results in pulmonary hypertension.
- Atrial septal defect (ASD, 8%): a hole between the upper chambers of the heart allows oxygenated blood to flow across the chambers and back to the lungs in addition to the normal flow. It also results in pulmonary hypertension.
- Patent ductus arteriosus (7%): this malformation is not related to the internal structure of the heart. A direct connection between the heart and the pulmonary

artery, which allows blood to bypass the lungs while in the womb, fails to close after birth and increases the amount of blood flowing to the lungs.

- Tetralogy of Fallot relates to four separate anatomical malformations, which together result in higher levels of unoxygenated blood being pumped around the body and is seen in 4% of those with a CHC (Seale & Shinebourne, 2004).

For those born with a complete AVSD, the recommended routine treatment is to have a surgical repair before the age of six months. This reduces the risk of post-operative complications including high blood pressure (hypertension). Mortality for this surgery is as low as 6% (Seale & Shinebourne, 2004). For other CHCs, patches or arterial banding can be used to prepare babies for future treatment surgery and surgical intervention, if required, typically takes place between the ages of 2 and 5 years, prior to beginning school (Backer, Mavroudis, Alboliras, & Zales, 1995). Post-surgery, children with Down syndrome are at increased risk of developing hypertension (Suzuki et al., 2000), however, they are less likely to need reoperation than children without a chromosomal abnormality (Seale & Shinebourne, 2004). Cardiac repair, in particular for AVSD, puts individuals at risk for residual problems including difficulties with blood flow, pulmonary vascular disease, cardiac arrhythmias, sudden cardiac death and bacterial inflammation of the heart valves. Cardiac repair will also have obvious implication in the form of time spent in hospital and gaps in education, but also may result in psychosocial outcomes noted in the typically developing population. Emotional reactions to chronic heart conditions include anger, anxiety, and depression that can contribute to social isolation, lack of physical inactivity, and generally withdrawing from activities that were once enjoyable (Falvo, 2005). The psychosocial impact of cardiac disease has not been monitored in individuals with Down syndrome and this has implications for surgery. For example, poor adjustment following treatment via an implantable cardioverter defibrillator (ICD) is routinely taken into account when deciding on the appropriateness of the treatment (Burke, Hallas, Clark-Carter, White, & Connelly, 2003).

In addition to congenital conditions, adults with an intellectual disability are at increased cardiovascular risk (Beange, McElduff, & W. Baker, 1995), and have a high rate of acquired heart disease. As discussed above, people with Down

syndrome are more likely to lead a sedentary lifestyle and poor diet, which will contribute to the development of cardiac conditions, although people with Down syndrome are reported to be somewhat protected from mortality that is due to coronary heart disease (Vis et al., 2009). People with Down syndrome are also at greater risk of hypothyroidism which in turn can suppress cardiac functioning; however, thyroxin treatment may improve development in children with Down syndrome and mild hypothyroidism (van Trotsenburg et al., 2005). Such risks related to a CHC that persist into adulthood indicate a greater need for specialist cardiac care for people with Down syndrome, specific measures to ensure they are able to access services and report symptoms and cardiac teams that understand their support needs and can assist decision making processes.

Within the population of people with a CHC, Down syndrome is a risk factor for hospitalisation (Kristensen et al., 2009). Prominent failures by services to provide good care, for example, in paediatric cardiac services such as at the Bristol Royal Infirmary, prompted the development of more stringent guidelines surrounding the process surgical procedures. As a direct result, the Parliamentary and Health Service Ombudsman for England published a detailed good practice guide to consent in cardiac surgery (Parliamentary and Health Service Ombudsman [PHSO], 2005). These guidelines outline the necessity for better communication, accurate reporting of risk, and clear procedures for keeping records when things go wrong. However, none of the four sites involved in the piloting of these guidelines reported the inclusion of people with a learning disability or their families. A subsequent report produced by the NHS (2007), signposts service providers towards a generic leaflet on consent for health care treatment written for people with a learning disability (DOH, 2001a). The report fails to provide any specific recommendations for staff training, treatment, or practice when someone with a learning disability requires cardiac surgery.

Health Provision for People with Down Syndrome

While individuals with Down syndrome are now living longer (Glasson et al., 2002; Janicki et al., 1999), a review of the literature indicated that people with Down syndrome may die up to 17 years earlier than their peers in the general population (Reilly, Hastings, Vaughan, & Huws, 2008) As people with Down syndrome and a

CHC experience higher mortality than those without a CHC (Frid et al., 1999), people with Down syndrome and a CHC may, therefore, not live as long as their typically developing peers. Despite this ongoing treatment for cardiac conditions is increasingly required into old age and services need to develop to keep up with the particular needs of their client groups. Perkins and Moran (2010) document the failure of health services in the USA to keep up with the aging needs of older adults with a learning disability, or provide learning disability or syndrome specific training to health professionals. There have been recent achievements in this area in the UK with much attention focused on the area of primary care and on the “need to find new and creative ways to support individuals to achieve their own optimum health and well-being” (DOH, 2009a, p.3). However, these guidelines include little information about how, or what, that support should look like or discuss the increased training of staff required to provide that support.

A recent report by Robertson and colleagues has investigated the outcome of the introduction of routine health checks for people with a learning disability (Robertson, H. Roberts, & Emerson, 2010). The systematic review identified papers from the UK and beyond and included reports of the medical outcomes of one-off health checks as well as studies where repeated health checks had been offered. They concluded that health checks are effective in identifying previously undetected conditions. In the UK, for example, between 94% and 51% of people included in studies had previously undiagnosed medical symptoms that required intervention (Baxter et al., 2006; Cassidy, Martin, Martin, & Roy, 2002; Wilson & Haire, 1990). Individuals with recognised medical needs such as cardiac conditions were found to have unmet symptoms including hypertension and high cholesterol (Wells, Turner, Martin, & Roy, 1997). Health checks resulted in further referrals for the majority of individuals, however, GPs did not always act on a referral, particularly if the screening had been carried out by a specialist health check service and not the GP (McConkey, Moore, & Marshall, 2002).

Few studies focused directly on the health gains that resulted from health checks and those that did largely relied on anecdotal evidence or examples including weight loss or successfully carried out interventions, such as ear wax treatments. For individuals followed across repeated health checks, referrals reduced over a five year period (Martin, 2003). Individuals with a learning disability and their carers

indicated that people “liked” seeing their doctor, and people largely thought annual health checks were a good idea (compared with six-monthly, biannual or three-yearly). Some individuals and carers reported fears such as a dislike of needles as a barrier to attendance (Robertson et al., 2010). Despite being offered health checks, there still remained a less than satisfactory uptake in some studies (Felce et al., 2008), and it is, therefore, also important to address the factors that may prevent people’s use of health services.

People with a learning disability report barriers to accessing health services and have unmet health needs; accessing community based services is problematic, and routine tests are frequently not arranged (Hayden, Kim, & DePaepe, 2005). They feel their views are not listened to and the learning disability label prevents access to better treatment (Edge, 2001; Lavery, Challis, Easters, Smitheringale, & Thompson, 2005). Parents of children with Down syndrome and a CHC have reported less favourable treatment for their children (Kmietowicz, 2001). Barriers to implementing or improving health screening reported by community learning disability nursing services included practicalities such as access to facilities and equipment; lack of skills and experience; time and insufficient support from management (McKenzie & Powell, 2004). GPs have also reported feeling unknowledgeable regarding the needs of their patients who have a learning disability (Phillips, Morrison, & Davis, 2004), GPs experience difficulty communicating (Cook & Lennox, 2000), and are unwilling to take responsibility for the health needs of their patients with a learning disability (McConkey et al., 2002). This reluctance to undertake health checks has been addressed by providing additional training for specific practices, developing a handbook about learning disability, and utilising GPs from other practices where necessary (Cook & Lennox, 2000; Perry, Felce, Bartley, & Tomlinson, 2010).

Health Research and Down Syndrome

Any planned improvements or changes to care provisions must include the recipients of that care in order to ensure it adequately addresses their needs (Bollard, 2003; Department of Health, 2001b). A literature search in Psycinfo, Cinahl, and Psycarticles databases, with ‘Down syndrome’ and ‘health’ as search terms results in over one thousand results. A narrowing of these results to the last 20

years (1991-2011) yielded 97 results once duplicates were removed. Topics covered by the research include epidemiological papers on oral health, physical health, and rates of Alzheimer's type dementia in individuals with Down syndrome; quantitative research involving parent reports of health service use, physical health, and oral health; a significant number of articles related to parent and professional views on pre-natal screening for Down syndrome; and a small number of qualitative papers asking parents their views on the physical health of their child with Down syndrome. Only two papers specifically focused on exploring the views of people with Down syndrome on the health services they received (Fender, Marsden, & Starr, 2007; Russell, 2006).

Fender and colleagues (2007) consulted older people with Down syndrome about what they wanted their doctor to do for them. Participants were able to indicate what they felt was appropriate for doctors to ask, what procedures doctors should carry out, and how doctors might gain information when people cannot or will not tell them what is wrong. Another project, (Russell, 2006), saw Down syndrome Scotland secure funding to run a three year health project using focus groups to involve people with Down syndrome in the development of accessible meaningful and appropriate health resources for people with a learning disability and their carers. Over the three year period, areas to be addressed were chosen according to common requests to the organisation and requests from focus group members. The focus groups also discussed the appropriate format for information and members explained they had little motivation to learn about pain or depression and, therefore, suggested that material on these topics instead be aimed at carers and family members. Group members were more motivated to learn about the positive aspects of health, including keeping healthy and self-esteem and learning more about health issues they were currently experiencing, including puberty or menstruation. Group members helped design colourful booklets for people with a learning disability on the topics of: getting older; dementia; diet and weight; death; keeping well; puberty; periods; and self-esteem related to Down syndrome. Booklets were produced for carers on identifying and supporting an affected person through: dementia, depression, puberty, pain, and coping with loss. Feedback received from group members, organisation members and the wider public indicate that the material was useful and the project worthwhile (Russell, 2006).

Research of this nature has also highlighted that poor prerequisite understanding of one's health can confound to create further confusion across time. McCarthy and Millard (2003) looked at the understanding of the menopause in women with learning disabilities. They found that many of them struggled to make sense of the menopause because they had never fully, or even partially, understood the meaning and significance of menstruation. In the same way, a lack of basic knowledge about cardiac conditions may prove a barrier to accessing treatment. Any intervention in this area should be in a form that matches the individual's ability and conceptual framework of the illness.

Why This is an Important Area to Explore

The research outlined above indicates that projects involving people with Down syndrome are both worthwhile and necessary. People with Down syndrome are living longer and, although the risks associated with CHC and Down syndrome have been widely reported, no clear guidelines exist to inform service users or service providers as to the specific needs of people with Down syndrome who receive care for a CHC. It is important that prevention and treatment are prioritised so that increased survival in Down syndrome can be matched by an increased quality of life (Bittles & Glasson, 2004). Bittles and Glasson go on to propose that increased life expectancy will generate "greater ethical and legal dilemmas in the treatment of people with Down syndrome, especially those identified as having the potential to benefit from organ transplantation and other major surgical procedures" (2004, p. 284). A multitude of factors are pertinent when making decisions regarding treatment including the potential outcome and long term quality of life of the individual. While currently social and economic factors are the main determinants of access to treatment, this could in time change to include individual preference on an equal access to all basis (Giraud-Saunders, 2009), and psychologists may have a key role to play in helping people with Down syndrome make decisions regarding their care.

For a long time psychologists have had a vital role within the NHS with regard to supporting members of vulnerable groups to make decisions (e.g. McCabe, 1996). Similar to children and adolescents, people with a learning disability may desire independent support when making medical decisions. In addition, where there is

reason to doubt the capacity of an individual to make an informed decision regarding treatment, a psychologist may be involved in an assessment of capacity under the Mental Capacity Act (BPS, 2006a). Clinical Psychologists working in NHS community learning disability teams may have long established relationships with individuals and, therefore, may be well positioned to monitor the consent and treatment process and ensure sufficient information and aftercare is provided. Without research as to what people with Down syndrome want from their cardiac care team, such a role may be difficult. Research as to any unmet treatment needs within this group will potentially have worthwhile outcomes for both people with Down syndrome and those working with them, including practicing psychologists. To provide guidance we must first understand the process and the available services – how do people feel about the care, information, and support they received with regard to a CHC? These questions must be answered by those affected and reflect their perceptions and needs rather than those of the service providers or researchers, which can often be the case in disability research (Hartley & Muhit, 2003), 2. Research has found that improvements in knowledge and self efficacy of staff treating people with learning disabilities leads to improvements in care (Melville et al., 2006), therefore, specific information in this area could improve access to care, and levels of care received.

Barriers to Research in the Area

A review of the literature has indicated a lack of previous research in this area. The NHS Community Care Act, 1990, initiated the need for consultation with service users in the assessing and planning of community services including healthcare (House of Commons, 1990). Valuing people (DOH, 2001b) and Valuing People Now (DOH, 2009b) set out targets for health and other services and note that services must “put the needs and wishes of the person using the service at the centre of their quality assurance systems” (DOH, 2001b, p90). Why then, are people with a learning disability not routinely consulted regarding their needs and wishes relating to the healthcare they receive? Population based research may not routinely or readily identify people with a learning disability (Linehan, Walsh, Van Schrojenstein Lantman-de Valk, Kerr, & Dawson, 2009), and there has been little

comparative health research that could highlight needs and barriers to care compared with other groups (Noonan-Walsh & McConkey, 2009).

There is a growing research literature that is attempting to involve people with a learning disability in research through participatory and, to a lesser extent, emancipatory research (Chappell, 2000; Nind, 2008). Methodological developments include the increased use of narrative methods and case study approaches to make research more accessible to people with a learning disability and also make research more receptive to moving beyond the tradition of articulating only individual or group experiences (Gilbert, 2004). Despite attempts to make research inclusive, Baxter and colleagues (2001) found that ongoing barriers to involvement in research, such as a lack of involvement in initial stages remains, and that increased time and financial resources are required to ensure inclusion at all stages. Additionally, although attempts continue to include people with a learning disability in research, they are not an homogenous group and people with Down syndrome have a particular pattern of health risks that makes it prudent to focus more specifically on this group (Jones, Hathaway, Gilhooley, Leech, & MacLeod, 2010; Smith, 2001). Within this group there are individual differences that may lead to the underestimation of the ability of people with Down syndrome to participate fully in research by those who are unfamiliar with them, including those responsible for granting ethical approval to carry out research.

Gaining appropriate ethical approval can create barriers to carrying out research in this area and, unless addressed, may reduce the likelihood that projects are developed. Paternalism presents a risk that decisions made by ethics committees result in the exclusion of people with a learning disability from research. Pettit (1992) reports about the development of a paternalistic stance via reactive dynamics to ensure past mistakes are not revisited (e.g. the Willowbrook study, Scanlon, 2007). Potentially increasingly conservative decisions by ethical committees, which are aimed at avoiding harm, also result in discrimination, as potential avenues for research are closed down. This may be caused by the misinterpretation of guidelines by multiple committee members who have a lack of specific experience of disability and may jeopardise research (Iacono, 2006). Disability organisation committees or committee members on general ethics panels are more likely to have a better understanding of the types and size of projects their

members are likely to benefit from. They are also best placed to ensure individuals are not inundated with requests to participate in research.

Particular considerations that apply to the inclusion of people with a learning disability in research include uncertainties regarding their capacity to consent, barriers created by living situations such as shared and supported accommodation, and a dependent relationship between the participant and the person conducting the research. Guidelines exist regarding the pertinence of the Mental Capacity Act (Department for Constitutional Affairs [DCA], 2005) in developing and carrying out clinical psychological treatment, research, and innovative treatment with people with a learning disability (BPS, 2006b) and the DOH has completed guidelines on supported decision making (DOH, 2007). Following these recommendations should reduce the risks to people taking part in the research and also the risk that ethics committees refuse to grant approval. Weighing up relative risks and benefits to research participation can be difficult and Dalton and McVilly (2004) suggest that individuals should be invited to participate based on the premise that “the research is related to the needs of people with intellectual disabilities and that the research has the potential to benefit people with learning disabilities” (Dalton & McVilly, 2004, p.62). In line with Participatory Action research (Santelli, Singer, DiVenere, Ginsberg, & Powers, 1998) and DOH guidelines (2007) in the present study, in instances where informed consent could not be provided, the wishes of the individual to participate in research were considered. Consent from a suitable other was considered to reduce as far as possible the risk that valuable stories go untold due to the reliance on verbal accounts. Attention was also given to the subtleties of the interview process and what goes unsaid (Booth & Booth, 1996). Booth and Booth stress the importance of “overcoming the barriers that impede the involvement of inarticulate subjects instead of highlighting the difficulties they present” (1996, p.67).

Summary

The available literature highlights the increased risks people with Down syndrome experience in relation to hospitalisation and mortality due to congenital cardiac conditions. Therefore, this is a specific area that warrants research interest. We know that despite improvements in the area of consent and cardiac procedures, the medical needs of people with a learning disability, which includes people with

Down syndrome, are not being addressed by the NHS. A wide range of people with a learning disability report multiple barriers to care and service providers report a lack of confidence when treating people with a learning disability. Research has found beneficial outcomes for staff training. Given the intersection of the dual diagnosis of Down syndrome and CHC, and its relative commonality for this group, improved knowledge could have important repercussions for care and support. Information surrounding optimum cardiac care should be gathered from people with Down syndrome and used to inform service delivery, staff training, and individual and family support.

The following research questions will be explored:

- What do people with Down syndrome understand about Down syndrome, their CHC, and the intersection of the two?
- What information have they received and what information would they like to have?

Chapter 2 – Methodology and Method¹

¹ The term methodology is used here refer to the theoretical analysis of the methods used rather than as a synonym i.e. the principles that determined how the method was initiated and put into action.

Participants.

Potential participants were adults who had a diagnosis of Down syndrome and a congenital heart condition (present since birth). Participants had to have sufficient verbal skills to take part in an interview. Five individuals completed interviews (Table 2.1). Four of these were recruited via a support group for people with Down syndrome and a congenital condition and their families (The Down's Heart Group, DHG). The fifth was recruited via a post on a learning disability email network. Once ethical approval had been granted, recruitment went smoothly and interviews were arranged to mutually fit with interviewer and participant schedules. All names and identifiable details have been changed. In line with previous recommendations aimed at ensuring participants felt at ease but also valued for their knowledge (Booth & Booth, 1996; Clarke, Lhussier, Minto, Gibb, & Perini, 2005), a collaborative approach was taken to the interviews, which were carried out as conversations between the researcher and the individual with Down syndrome. As part of this collaboration, participants were given the option of having other people present if they wished. Parents provided a varying degree of input into four of the five interviews. Simon was currently living at home as vulnerable adult proceedings had been instigated in his supported accommodation. His mother requested to be present for the entire interview and Simon agreed. Simon's father joined us for the second half of the interview with Simon's permission. Tom requested that his mother be present for the entire interview to help Tom and the researcher understand each other. Adam asked to complete the interview alone but his mother interrupted to offer us refreshments after thirty minutes. At this time I asked Adam's permission to clarify his health status as he had suggested his mother would be better able to provide information in this area. His mother answered my questions regarding his physical health diagnoses and then left the room. Jane and Katherine completed the interviews without parental support. Jane was not sure of the name of her CHC and I asked permission to confirm with her mother, who was present elsewhere in the house, after the interview. When parent views were provided the researcher gave participants an opportunity to comment in order to make it clear the participant's views were being valued also. Parent comments were included in the analysis as participants gave permission for their parents to speak for them and these comments are a reflection of the collaborative process behind the interviews. Interviews ranged in length from 37 minutes to 75 minutes ($M = 50.4$, $SD = 15.1$).

Table 2.1. *Participant Characteristics*

Name	Age	Ethnicity	Heart condition	Other health conditions	Living circumstances	Employment/ Education status	Parent/ carer present/ level of input
Tom (male)	19	White English	Cardiomyopathy, ICD defibrillator inserted	Hearing problems: has grommets	Living part-time with mother, step father and siblings	Attending a residential college	Mother present throughout, helped Tom and the researcher understand each other and gave considerable amount of additional information.
Katherine (female)	21	White English	AVSD	Asthma	With both parents	Attending college	No
Jane (female)	20	White English	AVSD	Overactive Thyroid, diagnosis of anxiety	With both parents	Unemployed, some voluntary work, seeking residential college place	No, mother consulted on name of heart condition only after the interview
Adam (male)	25	White Welsh	ASD, VSD, Patent ductus arteriosus	Visual problems, deaf in left ear, asthma, hypotonia	Supported accommodation 5 days, with both parents at weekends	Unemployed some voluntary work	Mother joined in middle and was consulted on specifics of health difficulties only
Simon (male)	26	White English	AVSD	Asthma	With both parents due to unsuitable supported accommodation	Unemployed	Mother present throughout, father present for second half of interview. Mother kept focused on interview and prompted some answers

Recruitment

The DHG was initially approached by the researcher to discuss the project. The group had been involved in previous research projects run by the researcher and was happy to facilitate recruitment for a project interviewing group members with Down syndrome and a cardiac condition, once ethical approval had been granted. Ethical approval was granted by the University of East London ethics committee. The DHG were holding a member conference in November 2010, and it was agreed that this would provide a suitable opportunity for individuals to volunteer for the project without being approached individually and, therefore, experiencing pressure to take part. The researcher spoke at the conference to explain the project and was approached by five individuals with Down syndrome and one parent whose son was not present at the conference, stating they would like to take part in an interview. At the same time information was also disseminated via a learning disability email network of which the researcher is a member. One parent responded stating she and her son would be happy to talk to the researcher. All interested parties were given or sent an information sheet (Appendix A), which was followed up with a phone call to ascertain willingness to take part and arrange a time and place to conduct the interview.

Rationale for Qualitative Research

In the current research, considering how people find meaning and make sense of their world was central. Qualitative research is a “situated activity that locates the observer in the world. It consists of a set of interpretative material practices that make the world visible. These practices transform the world” (Denzin & Lincoln, 2005, p.3). Qualitative methods provide a means of attempting to construct a psychological understanding of an experience and rejects the notion that some complex underlying phenomena will reveal an ultimate truth or rational, instead acknowledging the multiplicity and subjectivity of truths (Berger & Luckmann, 1967). Unlike quantitative research the goal was not to reduce the experiences of people to discrete relationships and predicted outcomes, but instead to open up a discussion to allow the pursuit of more, not less, routes of enquiry, therefore making qualitative methods most appropriate to this study. More specifically, and in line with Yardley’s writing (1997) on material discourses, the research sought to understand how the practical aspects of having Down syndrome and a heart condition feed into the identity, social relationships

and experiences of people with Down syndrome. By consulting people with Down syndrome on their experiences, while being mindful of any interpretative practices at work, this project aimed to provide a starting point to understand how it is to be a person with Down syndrome and a CHC.

Thematic Analysis

The recommendations of Braun and Clarke (2006) were used when planning and completing a thematic analysis of the interview data. The analysis aimed to provide a rich description of the data through the identification of patterns (themes) within the data and provide a description of these patterns alongside an interpretation of these patterns in light of the research topic (Boyatzis, 1998). Thematic analysis was chosen as most appropriate for the current data as the aim was not to create a detailed theory of the phenomena in question in the way an alternative method such as Grounded Theory might demand, nor would the phenomenological stance of Interpretative Phenomenological Analysis allow for the analysis of accounts aided by others (e.g. parents). Accounts may also have been lacking in the significant detail required to gain an in depth phenomenological understanding of the topic. Both Interpretative Phenomenological Analysis and Grounded Theory are frequently used to analyse interview data as they are forms of discourse analysis, however, the researcher's central aim was to stay as close to the sentiment and meaning of the original accounts as possible and avoid forcing a theoretical framework unto the potentially varied and oppositional experiences of individuals (Ussher, 1999). Through a critical realist position the thematic analysis aimed to note the ways individuals discussed and made sense of experiences, while also commenting on the impact of the broader social context on those meanings. While individuals with Down syndrome may be less able to contribute to research than more articulate subjects, this does not mean their experience is any less real, less influenced by social context, or less relevant (Rose, Thornicroft, & Slade, 2006).

Theme development.

Throughout detailed reading and re-reading of the data set, the researcher chose themes from patterns seen to repeat across the entire data set. This was a relatively small set of data and while themes that occurred more often were important,

more instances were not assumed to mean that that particular theme was more crucial. Themes eventually chosen were deemed to capture an element that was important to how individuals with Down syndrome and a CHC experienced healthcare and everyday life. The analysis was driven by the research title: What do people with Down syndrome and a congenital heart condition want from their cardiac team? Some participants verbalised more ideas and broader experiences than others and all opinions expressed were of equal importance, therefore, the themes identified are a reflection of the content of the whole set of data, rather than focusing on a detailed account of one aspect of the data (Braun & Clarke, 2006).

Themes were developed using an inductive/bottom-up approach, whereby the themes identified are linked closely with the interview data, rather than driven by the specific areas of interest of the researcher (deductive/ theoretical approach, Patton, 2002). This inductive approach focused on identifying semantic themes within the data to reflect patterns in the semantic content. These patterns were then interpreted and connected to possible meanings and implications through reference to the existing literature. This avoided moving beyond the data to concepts and ideologies, which although theoretically relevant, may have no resonance with the participants themselves, a pit-fall noted in previous research (McClimens, 2008). Despite attempts to develop an analysis that centres on participant views, the resulting analysis, as in any qualitative research, is a product of more than recorded interviews. It is influenced by the central research questions, the context of the research, the questions participants responded to in the interviews, and the assumptions that guided the coding and analysis of the interviews. Therefore, the analysis that follows is a product of the research process (Braun & Clarke, 2006), with the views of the interviewees at its centre.

Stages of Thematic Analysis.

Phase 1 - Familiarisation with the data.

The data was collected by a sole researcher, therefore, familiarisation with the data began prior to data collection and continued through the transcription process. Notes were kept during the interviews to aid transcription and make note of ideas for coding and further write-up. Once all five interviews had been completed transcription began. This method of not reviewing and analysing transcripts until all the interviews

had been completed ensured the process of analysis did not impact on the style or content of subsequent interviews. The process of transcription was slow due to frequent interruptions during interviews, over-talking, and difficulties understanding participants. Therefore, the transcription stage was valuable in becoming familiar with the data (Bird, 2005; Riessman, 1993). To reflect specific difficulties encountered, transcription conventions suggested by Bird (2005) were used. This helped to create a transcript useful for the research purposes (Lapadat & Lindsay, 1999), for example, including notes on tone and interruption. Once all five interviews had been transcribed they were printed landscape style on A4 paper. The interviews were double-spaced and line numbers were added to make it easier to identify quotes at later stages of the analysis. The interviews were read several times each and notes jotted in the margins to begin an initial list of all potentially interesting ideas contained in the data set.

Phase 2 - Generating initial codes.

The next stage involved the development of a long list of interesting features of the data (codes). The researcher began with the first interview and gave equal attention to each data item and interesting idea throughout all five interviews. These codes contained basic segments of information relevant to the topic in question. Coding was carried out first by hand, using coloured pens to differentiate from initial notes, and the interviews were read and coded multiple times to ensure all relevant data segments were included (e.g. Appendix B). Once all codes had been noted on the transcripts a list of these themes was created using a Microsoft excel spreadsheet, which catalogued the page number, line number, and the text from the quote itself for each occurrence of a theme. Some segments of data were coded into more than one code. This process resulted in a list of 500 code/quote pairs.

Phase 3 - Searching for themes.

This next stage involved analysing the codes to group them into potential themes. The database allowed the re-naming of codes to group them alphabetically into similar themes. These groups were then represented visually to identify how they related to each other using post it notes and spider diagrams (e.g. Appendix C). The researcher began to link codes through themes and different levels of themes. Some codes went on to create themes whereas others were combined to form other themes.

Phase 4 - Reviewing themes.

At this stage there were a list of six candidate themes and a collection of codes related to the process of interviewing that did not appear to fit with others (Appendix D). The candidates themes were reviewed in terms of their distinctness from each other and their internal coherence. At this point a theme centred on *health and healthcare* was reviewed and divided into two subthemes: “*Breathless and Dog-less*” and “*I do it by Myself*” to reflect the different focuses of health in a medical sense and health as a part of health and fitness. A third sub-theme – *different levels of treatment* was felt to fit better as a sub-level within the theme related to the self-care and getting support from others: *Self-Care, Care From Others and Gaps in Care*. The theme called *Down syndrome and identity* was reworked into a new theme, which combined is with “*Ask Mum*” into a new theme called *Who is the patient* to reflect the duality of both the individual and the mother as being in receipt of services from the heart team. On reviewing the entire data set, it was felt that some miscellaneous codes relating to the process of the interview warranted their own theme, given their representativeness of all interviews, and significant previous literature on the topic. This further theme called “*Don’t have a Scooby*” was created which included codes that related to the process of the interview, communicative confusion, and topic avoidance. As this theme was deemed to be related more to the process of research, the theme is discussed in Chapter 5 (Critical Reflections).

Phase 5 - Defining and naming themes.

At this stage, the themes were refined by clearly demarcating quotes that determined the essence of the theme. This process resulted in some changes as it became clearer what each theme was, and was not, about. Themes were kept simple and close to participant accounts. Where possible, quotes were used to name themes to keep the views of the participants at the forefront of the analysis. The theme was defined through the writing of a coherent narrative account of the theme, and any sub-themes, how the theme fit with the data set as whole, and how the theme related to the research questions and the wider literature.

Phase 6 - Producing the report.

The final product of the analytic procedure is seen in the Results and Discussion section below. Quotes from participants are used to demonstrate the prevalence of themes and allow the reader to decide whether each theme and

example are compelling illustrations of the story being told about the data. All names and potentially indentifying information has been changed to protect participant identity.

Ethics

Ethical approval.

Ethical approval was granted by the University of East London Ethics committee in August 2008 (Appendix E). The ethics application outlined the research questions outlined above and the methods that would be used which were informed by guidelines on carrying out research with people with a learning disability (Booth & Booth, 1996; Nind, 2008) Particular considerations included a single information and consent form in an accessible format (Mencap, 2002), building in opportunities to meet prior to commencing the research, and an ongoing process of consent in order to maximise the development of the therapeutic relationship and information gathered.

Consent.

Consent was sought to record each interview at the outset and detailed notes were made throughout to help the researcher with later transcription and also to provide reflexive notes on the research process. All participants consented to be recorded and the researcher's own Olympus™ digital recorder was used. Informed consent was addressed on an individual basis and aided by guidelines on consent produced by the BPS (2006b) and DOH (2007). Guidelines on the use of the Mental Capacity Act were also used (DCA, 2005). Consent was secured via a two stage process: verbal consent to take part was ascertained prior to enrolment and written consent was secured immediately prior to the commencement of data collection.

Ethical risk.

The process of research involves the negotiation of various ethical dilemmas. Firstly, challenges reported by researchers undertaking qualitative research include questions related to self disclosure, the impact of listening to previously untold stories, feelings of vulnerability and guilt, leaving the research relationship, and researcher exhaustion (Dickson-Swift, James, Kippen, & Liamputtong, 2007). Secondly, there may be difficulties with the research-therapy, and research-friendship, boundaries

(Dickson-Swift, James, Kippen, & Liamputtong, 2006). Although research interviews have the intention of being non-therapeutic, the interview may have therapeutic outcomes for both parties, or may create difficulties for trainee clinical psychologists not yet well versed in the management of such complex boundaries. Finally, research participation may inadvertently have negative effects despite attempts to avoid this and there may be risks to third parties mentioned in transcripts who have not consented to participate in research (Hadjistavropoulos & Smythe, 2001). Such risks were addressed in the current research by attending to professional guidelines (APA, 2002; BPS, 2006b), and research guidelines (Elliott, Fischer, & Rennie, 1999; Hadjistavropoulos & Smythe, 2001), including a clear explanation of the purpose and potential uses of quotes in material resulting from the research. After the interviews, participants were again reminded of the purpose of the study and given the opportunity to reflect on their feelings about the topic and taking part in the research, off-the-record.

Confidentiality.

In line with measures recommended by University ethics procedures, data was collected by the named researcher only and stored in accordance with the Data Protection Act of 1998, on a password protected external computer drive, stored in the researcher's home. All identifying data including consent forms were stored separately from interview recordings, demographic material and transcripts. Names and identifying details in interviews were changed and paper copies of interview transcripts were kept in a locked cabinet in the researcher's home. Each participant was assigned a research number and a list linking the pseudonyms of participants to these numbers was available to the researcher only and stored separately. Confidentiality measures were explained during the consent procedure (See consent form, Appendix F).

Reflexivity.

Undertaking qualitative research on sensitive topics with vulnerable groups requires reflection, imagination, careful preparation and trust on the part of the researcher (Dickson-Swift et al., 2007; Johnson & Clarke, 2003). Reflection was built into the design stages including careful consideration of decisions on issues including recruitment; informed consent; and assessment of risk. Throughout the process the

use of a reflective research journal (Andrews, 1996), and research supervision, helped reflect on the aims and purpose of the research and the role of the researcher. This ensured the methods used matched the needs of the project and the researcher was supported with decision making and the impact of research, for example, through supervision with a skilled research supervisor. Taking research days from placement allowed time to complete interviews and write detailed notes on the process of carrying out the research.

The assumptions and experiences of the researcher are particularly pertinent in qualitative research. The primary researcher in this project, a trainee clinical psychologist, had previously been awarded a PhD in learning disability. The PhD involved qualitative and quantitative research with families where a person with a learning disability had died. Many of these participants were bereaved parents of children with Down syndrome and a CHC. During this previous research the researcher was struck by the weight of responsibility placed on the shoulders of parents to make medical decisions and care for their children from the moment their child was born: 'We had to make a decision there and I remember saying, it's like putting us in the position of God' (Reilly, Huws, Hastings, & Vaughan, 2010, p.408). Such comments sparked an interest for the researcher in the views of adults with Down syndrome and a CHC and their experiences of decision making, hospitalisation, and healthcare. This previous research experience has had an impact on the current research. The prior research raised this as a potential suitable topic for investigation as part of the doctoral thesis. Prior knowledge of the literature and extensive contact with people with Down syndrome and their families through the previous project has had an impact on the development of the aims and focus of the project.

Also a working relationship with the Downs Heart Group provided a source of advice on the topics covered and questions asked. This prior knowledge of in the area combined with therapeutic skills gained during clinical psychology training ensure skills in finding the right balance to communication with people with Down syndrome and their families.

Advantage and Disadvantages of Research with People with a Learning Disability

Models of research with people with a learning disability have been developing over recent years following *participatory* and *emancipatory* routes. Participatory and emancipatory methods as applied in the area of learning disability have similarities in that they both aim to improve opportunities:

- for research to be lead by people with a disability
- for people with a disability to be involved as researchers
- for researchers to be more reflexive in relation to their work
- for greater understanding of knowledge of the difficulties and deprivations experienced by people with a disability and their family (Barnes, 2003; Iacono, 2006).

Emancipatory research goes further by insisting that researchers should be accountable to the democratic organisations of people with a disability and, to avoid conflicts of interest, these organisations should act as commissioners and funders of research (Chappell, 2000). Such calls for a change to methods used must be balanced with accessible and fair research and ethics protocols. Given ethics procedures and related barriers, and the fact that the health of people with Down syndrome with a CHC is an under researched area, it was felt that, instead, exploratory research using semi-structured interviews could be pertinent to identify whether further research and action in the area was warranted. Advocates were approached and consulted during the development phases in place of consulting people with Down syndrome themselves.

Interviews resulted in the collection of verbal and demographic data. The collection of verbal data limited involvement in the research to people with sufficient verbal skills to share their experiences and, therefore, the experiences of those at the more severe end of the learning disability spectrum will be missing. Such limitations have been similarly experienced by previous research (McCarthy & Millard, 2003). Booth and Booth (1994) outline arguments for and against the recording of interviews with “vulnerable participants”. It was decided that, as one of the main purposes of the research was to produce a piece of work to meet the requirements of a clinical psychology doctorate examination process, alternative methods of collecting data would be less suitable, such as depth interviewing involving repeated interviews with

the same people over a considerable period of time (Booth & Booth, 1994). Additionally, from the outset it was anticipated that we could have a small number of participants spread over a wide geographical area, therefore, ruling out the possibility of focus groups. It was also felt that audio recording of interviews gave flexibility with regard to time and location and reduced barriers that may have been posed by requiring participants to travel to a central venue. The importance of including people with a learning disability and or Down syndrome in research has been outlined in the introduction section. Throughout the project, decisions made kept potential benefits and risks to participants in mind (Dalton & McVilly, 2004).

Considerations of potential power imbalances throughout the project ensured that the methods used were inclusive and beneficial to people with Down syndrome. While qualitative research allows one to observe and study events in their natural settings, issues of power inherent to research have been well documented. Denzin and Lincoln (2005) explore issues of power within the development of qualitative research in a colonial context, whereby research reports generated from qualitative research were fundamental in developing strategies for controlling “the foreign, deviant or troublesome Other” (Denzin & Lincoln, 2005, p.3). This chapter has highlighted the decisions that were made throughout this research project including addressing issues of power, barriers faced, and the methods ultimately chosen to best explore and represent the views of people with Down syndrome and a congenital heart condition to a wider audience.

Interview design.

In order to access opinions on the topic, the interview was designed to address challenges in conducting research with individuals with a learning disability identified by Booth and Booth (1994): difficulties articulating views; unresponsiveness to open questions; difficulty thinking in abstract terms and generalising; acquiescence to yes/no questions; conceptual difficulties around time. Abstract language was therefore avoided, questions were direct, without orientation to time where possible, and time was taken to develop a trusting relationship with help from family members and carers where necessary. A semi-structured interview protocol (Appendix G) was developed, informed by previous research (Barter, Hastings, R. Williams, & Huws, 2010; Clarke et al., 2005). A list of potential questions was drawn up based on the available literature

on psychosocial implications of cardiac surgery (e.g. Arvidsson, Slinde, Hulthén, & Sunnegårdh, 2009; Stos et al., 2004), and discussed with the director of the DHG who advised on structure and potential prompts and alternative of asking questions (Appendix G) . Not all questions were asked but the schedule reflects the range of questions used to elicit information. The use of a structure was to ensure all planned areas were covered and to keep the interviewer on track. The interview included questions on lifestyle, medical history, experiences of primary and secondary care, symptom management, and plans and goals for the future. Findings in the literature indicate that parents of children with Down syndrome report that barriers to treatment were related to their child's condition. For example, they feel their views are not listened to and the learning disability label prevents access to better treatment (Lavery, Challis, Easters, Smitheringale, & Thompson, 2005). Parents of children with Down syndrome and a CHC have also reported less favourable treatment for their children (Kmietowicz, 2001). Therefore, participants were asked to tell the interviewer about themselves and, when all indicated that they had Down syndrome, were asked more about this to explore whether they saw a link between Down syndrome and their health experiences. The interview was generally participant lead rather than researcher or schedule lead. Communication styles and differing levels of ability required flexibility. People with Down syndrome are not a homogenous group and having fixed ideas as to the interviewing style required could have constrained the interview (Goodley, 1998).

Demographic details including name, address, age, gender, living situation, and employment were also collected from individuals. Additionally, details of the CHC and other medical history were collected. Parents aided this data collection process where necessary and permission to do this was sought from research participants. Demographic data was noted during the interview and was supplemented by questions at the end if any information was missing (Appendix H). This information was used to prompt further discussion and to provide context during the analysis process, for example, details of the CHC, current living and employment circumstances.

Epistemology

Epistemology is the study of how we come to know information, therefore, it influences methodological decisions, including the choice of method employed (Carter & Little, 2007). Research methodology considerations in the current project needed to take into account potential difficulties engaging people with Down syndrome in research and weigh any difficulties against the importance of gaining an understanding of the lives and needs of people with Down syndrome and, more importantly, the context within which research takes place.

Quantitative methods involve the reduction of material into discrete chunks of comparable information. In reducing information to clean, value-free forms there is a risk that the context could disappear completely (Banister, Burman, Parker, Taylor, & Tindall, 1994), or at least go unconsidered in terms of the exploration of results and relationships. In response to the proliferation of quantitative research methods, qualitative research has developed methods that take context into consideration, while exploring how social experiences are created and given meaning (Denzin & Lincoln, 2005). Qualitative research methods are utilised with the premise that the standpoint of the researcher cannot ever be value-free, as they require some initial knowledge of the existence of relationships to be researched and will always be situated within the political context wherein services are provided and received.

Multiple qualitative research methods exist and their development has been influenced by both realist and relativist views on how we get to know information. Positivist and post-positivist theory sees the world as a series of observable events that exist independently of human consciousness and cites rigorous scientific research as the only way to get to know, or at least estimate, this information. Some see this research as a suppression of the interpretative role of the researcher that takes scientific enquiry away from the real world in which events happen (Spindler & Spindler, 1992). There are researchers who accept such methods as one way of telling stories, however, alternative theories have grown in response to this purported suppression of interpretation. Realist theory agrees that the world is knowable but abstains from the detailed study of phenomena in order to uncover knowledge in a process that also takes into the account different perspectives, contexts, sources of power, and biases. Relativists agree with positivist theory that there is a world of observable events that exist independent of human consciousness, but state that our

knowledge of this world is socially constructed. For example, a social constructionist view “sees science as a form of knowledge which creates as well as describes the world” (Banister et al., 1994, p.9). The epistemological stance of critical realism adopted in the current study falls somewhere between these views.

Critical realists believe that “the task of empirical research is to explore how existing social, political and economic relations create inequality (...) in order to develop a normative critique against those relations” (Cruickshank, 2003, p.3). They acknowledge, however, that the researcher may not be able to access this knowledge directly and that reality will remain as it is regardless of the theories we have of it (Lopez, 2003, p.76). Critical realist theory allows for the examination of theory and concepts alongside the social and historical context which allowed them to develop, as this lets us question and update our theories of reality (Pilgrim & Bentall, 1999). The critical realist position has been chosen in light of the longstanding devaluation of people with Down syndrome by society (Nevel, 2010), and researchers (Iacono, 2006). Research has tended to be “about” people with Down syndrome from the perspective of parents and healthcare professionals, with the individual with Down syndrome seen as the “Other”, a problem to be dealt with and controlled (Denzin & Lincoln, 2005). The critical realist perspective acknowledges that historical and social factors have an impact on what we know about the lives of people with Down syndrome and, while exploring experiences from their perspectives may not change reality, exposing such social factors may provide added knowledge to theories on their reality.

Chapter 3 – Findings and Discussion

Following thematic analysis, codes and patterns identified in the interviews were grouped into three main themes: Who is the Patient, Self-Care, Care From Others and Gaps in Care and Health and Fitness (Table 3.1). Each theme is examined in turn, firstly exploring the meaning of the theme illustrated by quotes from interviews, then linking the theme to the literature and other available evidence in this arena. Particular consideration is given to assumptions of the researcher that contributed to the themes, links between themes and links to research in other areas. Implications these findings might have for people with Down syndrome, and those who support them, in light of the main research question, are explored in Chapter 4. When providing extracts from the interviews the following transcript conventions are used:

.. - Two second pause

... - Three second pause

(...) - Words omitted to shorten quote

[text] - Explanatory information included by author

I: - Interviewer

Table 3.1 *Themes*

1	Who is the patient
2	Self care, care from others and gaps in care
i	“I do it myself” – measures taken to manage health
ii	Help from others – the regular input of others to manage health difficulties.
iii	Emergency admissions and gaps in care
3	Health and fitness
i	“Breathless and dog-less” – implications of heart and health on fitness.
ii	“Its part of a strict diet”- eating for health.

Theme 1. Who is the Patient

This theme relates to identity: the ways in which participants described and represented themselves; what they felt was important to tell the interviewer; and what was most important to them and to the people central to their lives. In response to introductory questions about themselves and what they spent their days doing, participants talked about education and employment. Some were in education, others engaged in voluntary work and finding a paid job now or in the future was important:

I: Can you tell me what you do at the minute? Sam you're not at college, are you working?

Sam: Well Deirdre (interviewer) I have a few things that I do, well I do studies em I go on the laptop computer and em well I want to get a job.

Participants also explained their identity through stories of football teams, films they were fond of, youth groups and social outings.

Simon: And guess what Deirdre, in the college town I supporter of supported English Town

I: So you supported English Town

Simon: Yeah

I: So is football something you like?

S: Yeah

I: You like a lot of football teams

Simon: Yeah and I like music, play games and I real good at checking results and sometimes kick off, I'm really good

While some of these groups also involved people with Down syndrome and participants spoke of valuing friends who were similar to them.

I: Do you know anyone else with Down syndrome?

Adam: Ben

I: Is he a friend?

Adam: mm hmm

I: And what is he like?

Adam: Em....

I: Is he the same as you?

Adam: Uh huh

I: Is that good or bad?

Adam: Eh hmm [thumb up]

All participants were white-British, of a similar age, and constructed their identity in similar ways in terms of social networks, friendships and socially relevant activities such as further education, a move toward become independent (including financial independence) and gaining meaningful employment. Employment importantly meant an income:

I: So you might take her job off her [younger sister], and what you do there?

Jane: Same as her, be a sales assistant like sell.. em tags on clothes, paying on the till, taking the clothes when...[mimes]

I: Yeah when people bring them back

Jane: Yeah

I: That sounds like a good job

Jane: And it's a paid job [giggling]

I: A paid job, so is it important to have a paid job?

Jane: Yeah

I: So why is that important?

Jane: Get money to put in the bank. I've got quite a lot of money in the bank at the moment

Participants told stories of social occasions, school, charity work and accommodation. What seemed common to accounts were a reflection of three of the aims of the Valuing People White Paper: a desire for 'independence, choice, and inclusion' (p.23, DOH, 2001b), to make choices related to becoming independent including financial independence, being included in wider social events not just within the realm of learning disability, and making choices about where to live and, for example:

I: And what do you think you would like to do when you are older? Any ideas?

Jane: Em to have my own flat

I: Ooh that sounds good so have you started looking for a flat?

Jane: No not yet

I: Not yet but where do you think you might like to live?

Jane: Em somewhere not too far

I: Not too far from.. here?

Jane: Yeah like [inaudible] but on my own?

I: So why's that

Jane: I don't know really I just, I want my own flat, be independent

I: Okay so want your own flat so you can be independent

Jane: Yeah.

I: And what does being independent mean?

Jane: Em its means to have my own em house or whatever, flat just you know have my own space?

There were also examples of choices that could not be exercised because of health difficulties:

Jane: Yeah cause every year we have carnival up in the park and every year we have it and every year we have carnival and I missed two of them and I missed two, I missed my mates 20th and 21st birthday.

Choice and support to make decisions are concepts central to the Valuing People White Paper (DOH, 2001b). While health conditions may create obvious barriers to choice and inclusion, other barriers including people's beliefs around the ability of people with a learning disability to make a decision reduce available choices to a limited menu (Edge, 2001). The literature review above has outlined the lack of good practice guidelines related to consent in cardiac surgery for those with Down syndrome. The combination of higher mortality for people with Down syndrome and a CHC compared to people with Down syndrome without (Frid et al., 1999), a higher risk of hospitalisation (Kristensen et al., 2009), and a lack of evidence on ways of improving health in this group such as annual health checks (Robertson et al., 2010), will result in ongoing difficulties for this group. Difficulties such as making decisions that take into account their own wishes and choice, the implications of the health conditions and the views of healthcare professionals.

All participants knew about Down syndrome and generally, when asked, participants described Down syndrome as a “good thing”, however, some struggled to explain why they thought this:

I: So do you know about Down syndrome?

Adam: Errr..mmm

I: What can you tell me about Down syndrome?

Adam: Don't know

I: Is it a good thing or a bad thing?

Adam: [puts up thumbs]

I: A good thing! Two thumbs up.

Adam: mm hmm

I: So why is it good?

Adam: Emmm.. [sighs] hmmm.. don't know

Other discussions of the meaning of Down syndrome revealed feelings that Down syndrome was responsible for interviewees needing increased care and support from others. This was seen in both a positive and negative light. One woman reported that Down syndrome meant she was spoiled by her parents, which was good, as it would result in her parents looking after her in the future:

I: So what do you think about Down syndrome, is it a good thing? Is it a bad thing? Is it just in middle?

Katherine: A good thing

I: A good thing? So why is it a good thing?

Katherine: Because my Mum and Dad spoil me <laughs>

...

I: So it's good thing, just then you said it was a good thing cause Mum and Dad spoil you

Katherine: Yeah

I: Any other reason it's a good thing?

Katherine: Em my Mum takes me out shopping for clothes and my Mum takes to out to em the park and to the caff.

I: So they look after you because you have Down syndrome?

Katherine: Yeah

Worries about care in the future are commonly expressed within families of people with Down syndrome (e.g. Maxwell & Barr, 2003), and families of people with an unspecified learning disability have reported difficulties surrounding changes to care routines, such as the adolescent and early adult years (Todd & Jones, 2005). There is, however, limited literature asking people with Down syndrome about their future, a future significantly different to generations before given they are more likely to outlive their parents (Bittles & Glasson, 2004). Improvements to cardiac surgery and access to health care for people with Down syndrome have resulted in a significant increase to life expectancy (Janicki et al., 1999). However, other factors including deinstitutionalisation and improved living conditions are also related to reduced mortality (Noonan-Walsh & McConkey, 2009). In the future, changes to social care packages, increased available support and the introduction of individual budgets may replace the traditional methods of family support, however, such progress must also be balanced with considerations of safeguarding (Fyson, 2009). Risks associated with Down syndrome were recognised by participants, who explained that Down syndrome meant being more aware of vulnerabilities and people looking out for them in both a good way and in a way that was frustrating:

Jane: Em...um what you feel good about, its just a part of you that feels something special

I: Something special? So what's special?

Jane: Em cause like, I can get um, sometimes I get a bit vulnerable like when I'm taking money out of the bank unless like, could be in [names city] or anything

I: Yeah

Jane: Or I might get mugged or anything so..

I: So it makes you a bit vulnerable?

Jane: Yeah

I: So that's both.. maybe that's a bad thing sometimes, is it?

Jane: It's slightly

I: Little bit? Slightly bad?

Jane: Yeah

Planning for the future will require work with individuals, families and services to ensure that individuals continue to make choices related to their care without putting them at increased risk. Such considerations will include the knowledge that people with Down syndrome have about their health conditions, given the potentially dangerous implications of a poor perquisite knowledge of health conditions found by research in another area (McCarthy & Millard, 2003).

Interviewees spoke of the negative impacts of Down syndrome on their desires to have more independence in terms of accommodation and free time, wanting to have their own place and have more autonomy:

I: So what does Down syndrome mean? Is it a good thing or a bad thing?

Sam: Bad thing

I: Why is that?

Sam: Mum says do this do that [mimics growling orders]

I: So Mum tells you what to do?

Sam: Yep she push me

Jane, who recognised that Down syndrome put her at risk and made her “vulnerable”, lamented her lack of freedom:

I: Have your own space. Why would that be good?

Jane: I could have parties, get drunk [interviewer and Jane laugh] do naughty stuff

I: Can you not do that sort of stuff now?

Jane: No cause my Mum and Dad are here and they check me. It's bad isn't it so when you're with a boy

I: Oh okay

Jane: Not all the time it's just they won't leave me and my boyfriend alone.

This construction of identity also included a varied knowledge of the aetiology and potential impacts of Down syndrome. Jane and Katherine both mentioned the genetic process involved, whereas the others either did not answer the question or stated that they had no idea how it happened:

I: And how does Down syndrome happen?

Adam: I don't know

I: Is it something that you get when you are a baby or when you are older?

Adam: Em

I: What you think

Adam: Em

I: When you're a baby or older

Adam: Older

I: Do you know how it happens?

Adam: I don't know

There was also confusion as to whether others such as GPs and heart teams knew about Down syndrome and whether it was important for health professionals to know about Down syndrome:

I: So why do you think Dr Smith [GP] knows about Down syndrome?

Sam: Down syndrome means keep watch all the time

I: Really? What else does it mean?

Sam: It s mean em do as your told don't ask to watch on television em Mum and Dad say Sam is okeily dokey watch the wrestling now

I: And do you think that those doctors and nurses, did they know about Down syndrome?

Jane: Don't think so

I: You don't think so

Jane: No

I: So why do you say that?

Jane: They didn't, they didn't ask me ...

I: They didn't ask you anything about it, and what would you like to...do you think they need to know about Down syndrome?

Jane: They didn't ask, they didn't ask me any questions so, didn't get a chance [laughs]

While there was recognition that Down syndrome indirectly created some barriers to living alone and getting a job, participants saw that Down syndrome could have both positive and negative repercussions. There is a small and growing literature on positive perceptions of parents of children with a learning disability and

the impact that these positive perceptions can have on family life (Blacher & Baker, 2007; Greer, Grey, & McClean, 2006; Hastings & Taunt, 2002). The research literature has not yet included the impact of individual perceptions, whether positive or negative, on living with a learning disability or Down syndrome. The participants in this research did not blame the condition directly for their health and other difficulties and saw benefit in the difference it granted:

I: So what can you tell me about Downs? What do you know?

Jane: Well, em not much [laughs] you know. Em Mum was saying I have like, it's a chromosome like in my head, like actually in my body, doesn't my hands head or whatever

I: In everything?

Jane: Everything, and so I've got an extra chrom..., I've an extra chromosome, think it's like have a growth or something and its part of you..of me. I can't feel it [laughs]

I: No, it's just there, it's who you are

Jane: Yeah strange, natural

Jane was the most informed of interviewees, and was the only participant to allude to a connection between Down syndrome and health risks when talking about her anxieties around getting a swine flu jab. It was not clear, however, whether she was also considering the moderating impact of her heart condition:

I: How do you think you're going to cope with that? What are you going to do?

Jane: I'm not sure, I might be a bit rough after, for a few days so.. depends

I: But it will be good cause it will be better than getting swine flu?

Jane: Yeah...it might.. it might effect, em it might affect the heart and stuff.

I: Oh okay. So how might it affect your heart?

Jane: Well it might, I'm not saying that it will do

I: But it might do?

Jane: Might, its different, some people, you know when people do have Downs' em cause they will get affected won't they

Identity for this group did not focus on Down syndrome, but was more strongly focused on social networks and plans for the future. While parents reported and the researcher witnessed, an important role for mothers and fathers in supporting their children socially, this was not generally noted by interviewees. They tended to cite friends, teachers, and others as people they chose to spend time with socially. Defining oneself through social activity has been noted in previous research with people with a learning disability from a Jewish background (Bunning, Steel, & Science, 2006), and in a group of older adults with Down syndrome who had grown up in an institutional setting (Brown, Dodd, & Vetere, 2010). In this group participants may have also been reflecting the early adulthood stage they and their families were negotiating. Attempts to become autonomous could be viewed in terms of where they saw themselves as situated within the family life-cycle (Carter & McGoldrick, 1998):

I: And what do you think you would like to do when you are older? Any ideas?

Jane: Em to have my own flat

I: Ooh that sounds good so have you started looking for a flat?

Jane: No not yet

I: Not yet but where do you think you might like to live?

Jane: Em somewhere not too far

I: Not too far from.. here

Jane: Yeah like not too far but on my own

Alternatively, the creation of identity in this group may be fluid depending on the demands of the situation (Rapley, Kiernan, & Antaki, 1998). While a socially defined identity might be preferable, in situations where their characteristics as someone with Down syndrome or a patient with a CHC was more pertinent, participants were able to highlighted those parts of their identity. Todd and Shearn (1997) suggested that parental fears of labelling and stigmatising their children with a learning disability lead to a failure to disclose details of their disability to children. However, such behaviour was not evident in this group. Participants had a good level of understanding about their disability and reported that mothers had explained Down syndrome and cardiac information to them. When talking about Sam's level of understanding about his condition Sam's mother noted the difficulty she experienced

in knowing what and how much to explain and at what level to pitch information. Her comment could also be interpreted as a worry regarding potential stigma. She comments that she was “never a great one”, implying that she was never a great one for sharing information. This, in turn, suggests that at times she had held information back, although she had nonetheless explained Down syndrome and his CHC to her son:

Sam’s mother: Well yes he sort of well he does I, we tend to be open you know and em try and communicate the information to his level of understanding really its.. I do believe he knows a fair bit about. I mean the Down syndrome he has over em has friends with but I was never a great one... But he knows all these positive and he knows all these guys with Down syndrome and he can spot them and we went round one day and said you know have you got Down syndrome? And Sam said no and he laughed and said yes, so he does know

Whether they viewed their identity as positive or negative, participants embraced multiple parts to their identity. Not talking about their disability did not necessarily mean the disability was invisible to them or stigmatising (Beart, Hardy, & Buchan, 2005), interviewees may have been making a choice regarding that part of their identity and the prominence they wanted to give it given the research situation (Rapley, Kiernan, & Antaki, 1998). One part of the impact of defining one’s identity as someone with Down syndrome, or someone with a chronic condition, was that independence was sometimes curbed, and more support than desired was provided by others. The supporting role of mothers was experienced by the researcher. Three mothers and one couple met the researcher at the train station on arrival and in all cases mothers administrated the interviews. Two mothers attended interviews and played a role in steering questions and answers, providing advice on how best to pose questions and encouraged participants back on track when their attention wandered:

Adam’s Mother: [knocked then entered room] How you getting on?

I: Yeah doing okay

Adam’s Mother: Yeah, priming is the thing, you don’t always get closed, you have to have closed

I: Yeah

Adam's Mother: Use open questions and you'll get nowhere

There seemed to be a discrepancy between the independence and autonomy interviewees wished for and what they were allowed, or able to safely manage. Interviewees wished to be seen as a person first with the diagnosis of Down syndrome coming second. However, as we see in other themes, this was not the case in all areas of their lives.

Theme 2. Self-Care, Care From Others and Gaps in Care

In contrast to gaps in knowledge about Down syndrome and conflicting views of its impact on daily life, interviewees generally had a good understanding of their physical health, their CHC and agreed that the CHC placed limitations on their health and activity levels. This theme is divided into three levels: 1) "I do it myself" – measures taken to manage health; 2) Help from others to manage health difficulties; 3) Emergency admissions and gaps in care

(i) "I do it myself" – measures taken to manage health.

Individuals were generally responsible for at least aspects of their health on a day to day basis:

Sam's mother: What do we do? So we do it with the thing and what do we do once we've

Sam: Record the blood [over talking]

Sam's mother: yes record the number, the blood taken yes that's right

Sam: Give a squeeze the

Sam's mother: Mm hmm and you squeeze it out and what do you do? Where do you put it?

Sam: [over talking] that way

Sam's mother: That's right and where do you put it?

Sam: Monitor

Sam's mother: On the monitor that's right and what happens?

Sam: Check your blood

Sam's mother: And what does it tell you

Sam: You're point one

Adam, for example, looked after his ears by himself when in supported accommodation during the week, while his mother took care of this when he was at home:

I: So like Mum sprays your ears, will the staff spray your ears for you?

Adam: um

I: Or do you do it yourself?

Adam: I do it myself

Interviewees also talked about co-morbid conditions including asthma, hearing loss, allergies, and mental health difficulties. They played roles in managing these where relevant including knowing how and when to use an inhaler, taking daily medication, noticing physical symptoms, and seeking help from elsewhere when necessary:

I: So what if you weren't feeling very good. What if you felt [sniffs] a bit snotty or had a cough [coughs] what would you do?

Adam: [coughs] I got an inhaler in my bag

I: You've got an inhaler? So when do you take the inhaler?

Adam: Emmm... I don't know

I: So when do you.. you have an inhaler in your bag and what is that for?

Adam: Stop my bad cough

I: Stop your bad cough, okay, so can you show me? What do you do?

Adam: [mimes using inhaler]

They discussed the importance of visits to the GP, and indicated that GPs tended to provide support for less serious conditions including anxiety, menstrual pain, asthma, and weight loss. Interviewees were proud of their knowledge evidenced by the way they spoke and some demonstrating procedures, scars, and medication to the interviewer. This level of detail is similar to previous research with older people with Down syndrome that discussed the role of the GP and what doctors should and should not ask or be consulted about (Fender et al., 2007).

Fender and colleagues used group discussions in a series of meetings to explore what health meant for people with Down syndrome. As well as discussions about roles of healthcare professionals and lists of symptoms, participants linked health to fitness and activity as will be discussed in theme 3.

Managing one's own health also included knowing about healthcare professionals. In the current project interviewees reported that cardiac teams knew and understood about Down syndrome and spoke highly of their cardiac doctors and nurses:

I: Is there anything that you think the doctors don't know about Down syndrome or heart conditions or anything you'd like to tell them?

Katherine: Well my hero knows about my heart

I: Your hero knows so that's important

Katherine: Yeah

I: Anything else that you'd like to tell any of the doctors, that maybe they don't know?

Katherine: Em , Mr Smith, John [Surgeon] tells Mr Brown [Cardiologist] about my heart

(....)

I: And so why do you think they knew about Down syndrome?

Katherine: Because there's an extra gene inside me

I: So they understood about the extra gene

Katherine: Yes

The literature review has indicted that a CHC is a risk factor for hospitalisation in children with Down syndrome (Kristensen et al., 2009) and that people with Down syndrome are at a higher risk of co morbid physical health difficulties (Roizen & Patterson, 2003). Roizen and Patterson (2003) outline a health management approach that includes ongoing evaluation to identify changes in functioning, prevent of specific conditions, monitor ongoing conditions, and being vigilant for symptoms of specific conditions. The adults in this study seemed to be included in the management of their health. Ongoing assessment and monitoring and the maintenance of a good therapeutic alliance with their GP (Smith, 2001) are important factors in the maintenance of good physical health.

Alongside understanding and taking control of their own healthcare were difficulties that were out of participant's control. One difficulty common amongst interviewees who had been hospitalised since they had turned 18, was the challenge of staying on an adult ward, rather than on a paediatric ward. Problems noted by Tom and Jane, related to the difficulties of older people dying on wards, people with mental health problems on wards being disruptive, and a lack of activity available on adult wards.

I: And what was it like being in hospital?

Jane: Horrible

I: Why was it so horrible

Jane: Cause I end up, I got a bed, actually the first bed my first bed had like horrible rows like beds in rows..

I: In wards?

Jane: Yeah and em it was horrible cause em lots of ladies died next to me

Tom (aged 19), said that he preferred being on the "big boys ward", while his mother disagreed. Such a conflict raises questions as to how to rectify this situation. This issue is pertinent for any paediatric patients transitioning to adult care (Viner, 1999), but will have additional complications for people with Down syndrome, given the barriers they already experience accessing healthcare, including communication difficulties and behavioural problems (Hayden et al., 2005; Kmietowicz, 2001). These barriers indicate there may be limits to the level of healthcare people with Down syndrome can access without support and highlight the importance of consent procedures for this group and preparation for hospital admissions where possible (DOH, 2001a; PHSO, 2005). This issue may also require attention under the reasonable adjustments legislation. All NHS Trusts are required by law to carry out reasonable adjustments to remove barriers to care and ensure staff are adequately trained to provide care for people with a learning disability. A survey of these reasonable adjustments completed in Autumn 2010 by the Improving Health and Lives partnership (IHAL) found that while the majority of Trusts reported that adjustments were in place including accessible information, carer support, and specialist staff training, only a minority of Trusts were able to provide evidence of how these adjustments addressed the specific needs of patients with a learning

disability (Hatton, Roberts, & Baines, 2011). Trusts who reported on adjustments directly related to face-to-face contact with people with a learning disability most commonly relied on specialist staff training and liaison with Community Learning Disability Teams, however, the survey found a lack of detail regarding pathways to ensure adjustments were provided and maintained, particularly in times of limited resources. Importantly, few Trusts were able to provide specific details regarding the number of people with a learning disability accessing services, or numbers of those using independent mental capacity advocates (IMCAs). Better monitoring of adjustments could help develop guidelines in this area and ensure people with Down syndrome are not at risk on adult wards.

In terms of their understanding of treatment and hospital stays, interviewees varied with regard to their experiences of hospital treatment. For Sam, who had spent a considerable part of the last twelve months as an inpatient following surgery to replace two heart valves, questions about his heart condition largely elicited stories about this recent hospital stay. He was able to make clear links between the time in hospital and his health. Although he had been frustrated by his time in hospital, he recognised that the treatment he received had been necessary to help him gain the strength to walk:

I: Were they nice or were they nasty?

Sam: Nice

I: They were nice, what did they do?

Sam: Make me walk

I: They made you walk., which is good, what else

Sam: They would say catch the ball then push chairs, wheelchairs go [makes wheel sounds] oh go go go go down corridor hurry up hurry up hurry up hurry up hurry up [laughs]

Other interviewees talked about being 'poorly' as children and some articulated accounts of emergency surgery and parents being upset when they were born, due to their ill health. While knowledge of the functions of surgery was mixed, there was good understanding that this had happened a long time in the past:

Katherine: I was really poorly because I couldn't breathe myself and my Dad was crying

I: So you couldn't breathe and you cried a lot [misheard]

Katherine: I couldn't breathe and my Dad was crying

I: Your Dad would cry awh, so why was Dad crying?

Katherine: Er I think they thought I was gonna die but i'm not

I: No you didn't, so Mum and Dad were worried you were going to die, that must have been hard for them

Katherine: Yeah, I came out fit and healthy

While interviewees recognised that they had overcome ill health in early life, they connected ongoing health difficulties to the heart condition and were able to explain medication routines and medical terms and procedures:

I: And do they ever give you injections?

Adam: mm hmm

I: And what's that like?

Adam: Have blood taken

I: Oh so they take your blood?

Adam: Yeah

I: Ah and why do they do that?

Adam: That's why I'm very sick that's why.

All provided valid reasons for hospital check-ups and ongoing contact with cardiac teams such as current ill health, checking everything is okay, and risks to health due to their heart condition. Interviewees spoke positively about cardiac team members, linking the role they had in previous surgeries to their current monitoring of health:

I: So they are heart doctors?

Tom: They stick some things unto me

I: so they stick things on and what are they for?

Tom: Listen for my heart

In summary participants were able to take responsibility for their health, and physical symptoms, and had a significant knowledge of their health status and the cardiac team involved in their care. This information could, in theory, contribute to consenting to a medical procedure or making a decision about treatment, even if

their understanding is deemed insufficient for individuals to make the decision by themselves. The Mental Capacity Act (DCA, 2005), indicates that it should be assumed that an individual has the capacity to make a decision unless it can be shown that capacity is lacking at the point where a decision is to be made. In addition the individual must not be assumed to lack capacity just because the decision is an unwise one. Individuals who have experienced a lifetime of physical health interventions may choose to refuse a treatment. With this in mind when someone is deemed to lack capacity and decision made in their best interests by others must be an option that is least restrictive of their rights and freedom (DCA, 2005). For those deemed capable of making their own decisions, the Department of Health encourages supported decision making in practice (DOH, 2007; Hardy 2010). There is a need to develop an evidence base on the implementation of guidance related to the provision of support for people with a learning disability who are making medical decisions, as there is a risk that too rigid a focus on 'choice' could be dangerous if these decisions are beyond the abilities of people with a learning disability (Flynn, Keywood, & Fovargue, 2003).

(ii) Help from others – the regular input of others to manage health difficulties.

Tom's mother: what else? What else do you take if you have pain?

Tom: Mum

Tom's mother: Yeah you have Mum but what else do you have?

Tom: Spray

While all interviewees spoke about the things they could do for themselves, all accounts also included details on the roles mothers played in supporting individuals with treatment, in appointments, and co-ordinating their healthcare. Such accounts highlighted that in terms of treatment by medical teams and GPs, people with Down syndrome were very much supported by family members and mothers were highly involved in care and decision making. At different points in all interviews participants reported limits to their knowledge, such as not knowing the name of their heart condition, not knowing how it happened, or not knowing precise details regarding the functions of medications. Interviewees directed the researcher to

mothers for further information, or indicated that this was a matter for the doctors, whether cardiac teams or GPs. Interviewees also told of the support mothers provided and, while there was pride to be felt in accounts of taking responsibility for one's own health, there was nothing more than a nervous laugh to indicate embarrassment or anger at having to rely on others for help:

I: And what's the doctor like when you go there?

Katherine: Nice

I: Nice? Do you go by yourself or does Mum come in with you?

Katherine: My Mum.. and my nan

(...)

I: and who does the talking? Do you do the talking or does Mum do the talking?

Katherine: Mum [laughs]

I: Does she? You are well able to talk for yourself

Katherine: Well sometimes I do sometimes I don't

I: So when you don't, why is that? When you don't do the talking?

Katherine: Cause I'm nervous

The involvement of parents raises issues of consent and highlights that, as noted above; at times people will need support to make decisions or may be unable to make decisions (DCA, 2005). Mothers noted ways they managed issues of consent through negotiating relationships with professionals, however, there was no evidence of the guidelines for consent being pursued by professionals (NHS, 2007; PHSO, 2005). Accounts included no detail of parents and professionals being explicit about the decisions they were making, as is illustrated by Sam's mother who explains a recent conversation with a nurse when her son was attending for a flu jab:

Sam's mother:...whereas she he did say no and we said yes and she said I can't do it and we said yes you can we have an agreement with Dr Smith [GP]. 'Ah fine' [said in high voice to impersonate nurse]. So when she sort of got through that initial she, I understood where she was coming from and we said but we've got an arrangement, we've got an arrangement with Dr Smith and she said okay that's fine then.

Some interviewees expressed preferences for doctors talking to mothers or having the support of mothers for checkups and trips to the GP:

I: So sometimes they talk to Mum more than they talk to you?

Tom: Er yeah

I: and what do you think of that. Is that good or bad?

Tom: Good

I: Good so you like it when they talk to Mum?

Tom: uh huh

I: What would be better, if they talk to Mum or talk to you? Which would be better?

Tom's mother: Do you like them talking to Mum or do you like them talking to Tom?

Tom: To you [points at mother]

Tom's mother: To me?

Tom: Yeah

While procedures for consent may have been present, but not explicit, there remain concerns about those who do not have parents to support, in terms of how much they understand about treatment and who supports them with treatment. An incident in a supported living house had resulted in Sam refusing important medical treatment:

Sam's mother: The last house scare he em said no to taking his meds and he got away with lots of things

Parental accommodation to and negotiation of the world of learning disability services has been noted to take time (Todd & Shearn, 1996), and parents report less reliance on services over time due to service limitations. Parents have also reported that interactions with services can involve conflict (Todd & Jones, 2003) and have experienced barriers accessing services for their child with Down syndrome and a CHC (Kmietowicz, 2001). Raising a child with Down syndrome requires resilience on the part of parents in the face of change and challenges (Van Riper, 2007). Social systems theory has been used to explain coping and responses in families of people

with a learning disability, in particular with reference to the sharing of information and resources (Grant & Ramcharan, 2001). Better understanding of social systems, and involving parents in planning healthcare services, could improve relationships and adherence to consent procedures. However, this should not exclude the individual with a learning disability and the views of parents and their children may differ. The Mental Capacity Act indicates that any major medical decisions should include consultation with an Independent Mental Capacity Advisor (IMCA) (DCA, 2005).

(iii) Emergency admissions and gaps in care

Mothers present for interviews noted that cardiac teams were “pretty good” when it came to communicating with patients and families, but this changed when hospital admissions were unscheduled. On occasions where admissions were necessary to local hospitals or A&E departments, difficulties occurred including food allergies and unskilled staff:

I: So was it...what was the food like? Did they give you okay food?

Tom: Umm yeah not nice. They gave me baked beans

Tom's mother: Yeah that's right. He's allergic to baked beans and they served him some baked beans

I: Ok oh dear

Tom: by self

I: so that wasn't good. That was a bad thing wasn't it?

Tom: Yeah

Also families differed with regard to contact with their local GP. Tom's mother explained that their GP provided little support and tended to panic when advice was sought. Sam's mother reported a good relationship with the current GP, but explained they had problems in the past with an older GP who knew little about Down syndrome:

Sam's mother: He did have a problem, we did have problems at the beginning cause he had an older GP who was.. he clearly had a chest infection because I mean his un-operated heart defect.. and the older GP said give him orange juice rather than antibiotics. So we used to end up in the hospital, in the local hospital.

While care within teams who routinely had contact with participants was largely satisfactory, input from emergency teams, older GPs and less specialist services such as local hospitals was less so. Much has been invested to improve access to health services and services received by people with a learning disability, such as implementing annual health checks for people with a learning disability across the UK (Emerson et al., 2010), however, these checks will largely be beneficial for those not already in contact with physical health service providers. Additionally the legislation regarding “reasonable adjustments” outlined above will be relevant when people with a learning disability are admitted to a medical setting in an emergency situation. Adjustments made in emergency situations identified by Hatton and colleagues included the use of ‘flags’ to indicate that the patient with a learning disability had additional needs (Hatton et al., 2011). Trusts also reported the use of communication tools such as health passports created and maintained prior to admissions, which are then available in the event of an admission, planned or otherwise. The survey included an evaluation of two ambulance trusts who provided no information on the adjustments they provide in the event of an individual with a learning disability using their service. The results indicated that people with a learning disability continue to experience inequalities accessing healthcare. This is in line with the findings of the Michael Inquiry (Michael, 2008).

In the current research, interviewees and parents mentioned specific team members who were skilled in communicating with them or their child, however, in situations where staff were less familiar with the families, such as A&E and local hospitals, adjustments were not readily evident. Tom and Jane both reported they were given a side room after a number of days on an adult ward:

I: Two weeks in hospital wasn't nice?

Jane: No, I moved to em a side room too

I: Okay

Jane: After a quieter ward to side room, oh nice [puts on voice to impersonate nurse]

I: Okay so why did you move to a side room?

Jane: It was too noise, it was too noisy

I: So was it better in a side room?

Jane: Yeah

Side rooms also facilitated parents staying overnight, however, making adjustments to allow parents stay in hospital is not an adjustment for the individual themselves, nor does it solve difficulties with the lack of staff expertise in providing care for adults with Down syndrome. The IHAL survey (Hatton et al., 2011), also reports that the majority of Trusts said that accessible literature was available from Trust intranets or on request and only 9% (8 Trusts), indicated that easy read information was routinely available in wards and departments. This suggests that information is not available when admissions are unplanned, which is a reflection of the experiences of interviewees who experienced emergency admissions as less satisfactory than routine treatment.

In summary, interviewees were capable of managing their own health to a point and had knowledge of certain procedures and medications. They also sought the support of their mothers when required, indicating that conversations around consent to procedures are important for this group. However, adult wards, unplanned care, and emergency admissions posed problems to participants and staff unskilled in working with people with a learning disability were making admissions unsatisfactory through difficulties with communication and at times participants were put in danger (e.g. food allergies). A Department of Health inquiry has previously identified barriers that people with a learning disability experience when attempting to access NHS services (Michael, 2008) and the current research indicates that while there was evidence of good care, people continue to experience inequalities and report negative experiences of healthcare. The accounts indicated that “reasonable adjustments” (Hatton et al., 2011) being made to improve hospital experiences, such as moving people to side rooms. Accounts also indicated, however, that individuals continue to face risks during hospital admissions including exposure to allergens and that there is work to be done before they receive a service on a par with that received by patients without a learning disability.

Theme 3. Health and Fitness

The third theme relates to health in its broader sense within the realm of health and fitness. This theme is broken into two levels: 1) “Breathless and dog-less”- implications of heart and health on fitness and 2) “It’s part of a strict diet”- eating for health.

(i) “Breathless and dog-less” – implications of heart and health on fitness.

All participants talked about the importance of exercise and keeping fit. This was in light of ongoing health problems and interviewees, generally, had a good understanding of the limitations placed on their health by their heart conditions:

I: And what were they for? .. What did they help with, the inhalers?

Sam: My breathless, dog-less

I: So they helped with being breathless

Sam: And dog-less

I: And dog-less, So they help you walk your dog

Sam: Yeah, exactly

Interviewees discussed having to take breaks during physical exercise, monitoring and responding to physical symptoms, and taking preventative medication.

I: So do you have to learn to notice when you're too hot [mimes]. So you think I'm too hot I have to sit down.

Tom: yeah drink apple, apple juice

I: So you have a drink as well? That is important

Tom's mother: What else? What else do you take if you have pain?

Tom: Mum

Tom's mother: Yeah you have Mum but what else do you have?

Tom: Spray

Tom's mothers: You have your spray

I: Ooh

Tom: So you do that and on your tongue like that [mimes action of oral spray under tongue]

In reaction to challenges to health and fitness, participants spoke of the importance of keeping fit and healthy for both specific and general health benefits. Advice regarding fitness frequently came from GPs and interviewees were involved in multiple fitness activities including going to the gym, dance classes, horse riding, swimming, walking, and dog walking:

I: You're gonna lose weight? Tell me about that

Adam: [slaps stomach and flexes bicep on right arm]

I: [laughs] So you're going to be a muscle man?

Adam: Yeah! [emphasises]

I: And how are you going to do that?

Adam: [flexes biceps on both arms]

I: That's some big muscles

Adam: Mm

I: So what are you doing to get big muscles?

Adam: Em a bike ride

I: Bike ride what else?

Adam: Gym

I: Gym, what do you do in the gym?

Adam: Em I [sharp intake of breath, mimes weight lifting]

I: So you lift some weights

Adam: Mm hmm

I: What else?

A: Swimming

Sam, in particular noted the paradox that his health stopped him having the energy to exercise and walk his dog, however, increasing activity helped prevent chest problems:

I: So if you are at the gym and you get a bad chest

Sam: um yeah

I: what would you do

Sam: I meant that.. go to the gym and lots of walking fix up the chest

I: So they help with your chest?

Sam: Yeah

What became apparent in all interviews was the importance of support to get fit and access fitness facilities. Interviewees went to the gym with friends and family members and spoke of parents and others encouraging them to get fit and healthy:

Katherine: And they have the stepping thing... my Mum does that, she wants me to go on it, where you stay in shape.

I: So do you go with your Mum?

Katherine: And my Dad

I: You all go together?

Katherine: Yeah

I: Who takes you swimming?

Adam: My Mum and my staff

This role of families and friends in improving health and fitness is important in light of research documenting the so called 'sedentary' lifestyle of people with Down syndrome. The increased physical health risks faced by people with Down syndrome have been documented above. A sedate lifestyle and poor diet contribute to physical health difficulties including diabetes mellitus, arthritis, and obesity (Braunschweig et al., 2004; Roizen & Patterson, 2003). In addition, people with Down syndrome are at a greater risk of developing conditions, which in turn limit physical activity as they age, such as osteoporosis and vision problems (Barnhart & Connolly, 2007). High levels of inactivity (Draheim, Williams, & McCubbin, 2002), and lack of access to physical training programs (Temple & Walkley, 2007), have been reported for people with a learning disability generally. Physical training programmes must also be developed with the needs of people with Down syndrome in mind and a life-long approach may be required to beat obesity (Roizen & Patterson, 2003).

While this relatively young group of participants were well supported to access physical activities this may not be the case for all adults with Down syndrome, particularly as they get older. People with Down syndrome can struggle to maintain friendships once they leave the education system (DHaem, 2008), and informal support may not be as readily available as parents, and individuals, age. Skill acquisition through education and parent support are, therefore, important to consider. Lower body mass index in adults with Down syndrome has been found to be correlated with increased access to social and leisure activities and higher reported satisfaction with friendships (Fujiura et al., 1997). This relationship is unlikely to be straight forward but it is possible that supporting people to maintain

good levels of fitness will have implications for their social lives, and that supporting people in their social lives may also have a positive impact on their health.

(ii) “Its part of a strict diet”- eating for health.

Alongside this knowledge of the value of exercise was an awareness of diet and weight loss and the link between exercise and being a healthy weight:

I: Oh So the gym and what does the gym do?

Sam: Em well burns calories

I: Burns calories? So why is that important?

Sam: I use my phone and tell me how much I use

I: And what does burning calories do?

Sam: It's a Nokia

I: Does it help you lose....

S: Weight

This second level encompasses knowledge of weight loss, motives for weight loss, and strategies for weight loss (exercise and diet) and indicates ongoing interventions to manage and prevent obesity (Roizen & Patterson, 2003). This topic was generally raised as part of a discussion about communication with doctors, as the perception was that doctors and teams have an interest in weight and encouraged participants toward exercise and dieting:

Katherine: The doctor says [changes voice to impersonate] “well she's doing okay at the moment she's on a diet, she needs to go on a diet to lose a lot of weight” which is very hard to do

Motivations for changing eating habits included weight loss, improving muscle strength, and maintaining a slim appearance:

I: And what does eating them mean? Does it make you strong?

Adam: Like Gaston on Beauty and the Beast [Disney Film]

I: Oh so you want to be strong like Gaston. And what's good about Gaston?

Adam: He's like eggs, make him muscles.

I: So he's got muscles. And why is muscle good?

Adam: Em... I lose weight

Strategies for weight loss included cutting out junk food, eating less and eating smaller portions, eating lean foods, eating more fruit and vegetables, and drinking water. Participants talked about taking food education or cookery classes at college and also learning to cook through watching television programmes with celebrity chefs such as Jamie Oliver:

Sam: Mum says hurry up it Jamie Oliver hurry up its Jamie Oliver I'm coming.

I: So you watch Jamie Oliver on telly

Sam: Yep

(....)

I: And do you learn to cook with Jamie Oliver as well?

Sam: Yep

I: So what have you learned to cook

Sam: By follow the steps

I: Follow the steps?

Sam: and instructions

For one interviewee, fitness and exercise were linked mainly to muscle development and he spoke of the different sweets and chocolate he spent his money on, suggesting less of a focus on healthy eating. Individual accounts of advice suggested to the researcher that advice had been given in a way that felt supportive and could be utilised by those interviewed. Interviewees recognised that fitness and weight loss were hard work but they were committed nonetheless:

I: And is it hard to be on a diet or is it easy to keep to?

Sam: It is, its part of a strict diet

I: Strict diet

Sam: Yeah mm hmm

Mothers again provided support with cooking and dietary advice and this has implications for those who do not live with families, or who strive for greater independence. Participatory interviews with young people with Down syndrome and their families have previously found poor knowledge of healthy food and exercise and basic hygiene was not maintained without parental monitoring (Jobling & Cuskelly, 2006). Approaches to supported living such as Active Support (Mansell,

Beadle-Brown, & Ashman, 2010), aim to make everyday skills, such as cooking, accessible and available to all, including those with severe and profound learning disabilities and behaviour that challenges. However, evaluation of programmes suggests that cost and pressures on staff time can create barriers to implementing support and that choice with regard to support needs to be addressed (Standcliffe, Jones, Mansell, & Lowe, 2008). Making healthy eating and cookery accessible to all is imperative if health improvements are to be long term.

For the interviewees, health and fitness was more than taking tablets and avoiding risk. The interviews highlight that a multi-faceted approach to being healthy, including adherence to treatment regimes, consultation with healthcare professionals, taking responsibility for one's own health, maintaining levels of fitness, learning about nutrition, and implementing this knowledge, is achievable by people with Down syndrome. This highlights the need to focus on the reduction of barriers typically faced by people with a learning disability getting involved in physical activity (Barnhart & Connolly, 2007; Rimmer, Riley, Wang, Rauworth, & Jurkowski, 2004). Economic factors and physical barriers in the natural environment present major barriers as do individual emotional and psychological factors (Rimmer et al., 2004). A high level of support will be required, and family members and healthcare professionals will also require education and support. Where family members are not available, higher levels of monitoring may be required to ensure information is provided, understood and implemented. Learning disability services frequently experience high rates of staff turnover, therefore, procedures need to be developed to ensure a consistent approach is implemented (Mansell, Beadle-Brown, Macdonald, & Ashman, 2003).

The analysis highlighted the depth, and also the variety, in levels of knowledge and involvement in self-care for cardiac conditions. Theme 1 – Who is the Patient, outlined how people with Down syndrome and a congenital heart condition view their medical condition. It highlighted the integration of ability, disability, and goals for the future. While Down syndrome itself is an underlying cause of a CHC, it was not blamed for medical difficulties. At times it resulted in limited independence for individuals, which was perceived as both a positive and negative outcome. Theme 2 – Self Care, Care from Others and Gaps in Care identified the roles people took in managing their own health, whilst also wanting appropriate support from

parents and healthcare professionals. It was also noted that there were gaps in services, which individuals wanted filled, such as better communication and support while in hospital. Theme 3 – Health and Fitness, explored the important link between fitness and health as explained by people with Down syndrome and a CHC. Individuals were supported in this by parents and healthcare and social care staff and had a good understanding of the importance of keeping fit in order to stay healthy, and keep one's heart healthy. Measures taken to maintain health and fitness indicated evidence of ongoing interventions to counter the historical poor health and sedentary life style of people with Down syndrome.

Chapter 4. Implications and Limitations

In response to the research question: What do people with Down syndrome and a congenital heart condition want from their cardiac team, the findings outlined above indicate that the people with Down syndrome and a congenital heart condition want a good relationship with a cardiac team who understand Down syndrome, understand the additional limitations Down syndrome might pose and be prepared to support both them and their families through treatment across the lifespan. Participants involved in this research had a good understanding of both their cardiac condition and their diagnosis of Down syndrome. They outlined the limitations posed by both conditions and talked about personal goals, which have important implications for treatment including striving for independence and employment. Participants discussed the efforts they made to look after their health, but also the help that they sought and welcomed from parents and healthcare professionals. Participants were able to identify different roles for cardiac team members and GPs, for example. There were times when participants preferred healthcare professionals to talk to their parents rather than to them, which raised the important issue of consent to treatment and supported decision making. Participants, additionally, explained the important link between health and fitness, reporting on various sources of support with their fitness and also making it clear that, while fitness was difficult to achieve, it was important for multiple reasons. This chapter will outline the implications of these findings, while also examining limitations and suggestions for improvements and further avenues for enquiry.

Implications

These findings tell us that people with Down syndrome and a congenital heart condition have valuable accounts of their experiences to share, which have not yet been incorporated into the wider literature or service development, despite the wider recognition of the importance of service user input into service development for the future of healthcare services (DOH, 2006b). People with Down syndrome and a congenital heart condition are living longer, with conditions not previously seen in later life, and a greater range of treatment options will result in new and challenging decisions to be made (Bittles & Glasson, 2004). The role of parents in supporting medical treatment and decision making creates a potentially complex situation, whereby people with Down syndrome and their parents may have differing views of

what treatment and lifestyle choices might be best. The Mental Capacity Act (DCA, 2005), provides a clear protocol for supporting difficult and potentially unwise decisions and as people with Down syndrome grow older and attempt to assert their independence and choice, decisions will need to be carefully supported and monitored over time (Edge, 2001). The provision of independent advocates outlined by the Mental Capacity Act (DCA, 2005) for those facing serious medical treatment is useful in principle, in practice concerns has been raised that a lack of skills in practitioners using the act may limit appropriate referrals and in turn limit appropriate support for those lacking capacity to make decisions (Redley, Luke, Keeley, Clare, & Holland, 2006; Sawhney, Mukhopadhyay, & Karki, 2009). With regard to medical decisions in particular, medical professionals have been found to have a negative view of the potential contribution an IMCA could make to medical care (Luke, Redley, Clare, & Holland, 2008). The current findings could improve medical teams' understand of the needs of their patients. Decisions regarding medical care should not be based on medical outcomes alone, but must also take into account the priorities and goals of the individual and the person who is best positioned to support them in that decision, regardless of their medical training.

The current research has important implications for healthcare services and clinical psychologists who work in teams that provide medical treatment to people with Down syndrome. This study allowed people with Down syndrome to have their say on the services they receive and illustrate what is important to them in terms of how services are delivered. As in previous research, participants were able to provide feedback on services and also give insight into their understanding of their health and care (Bollard, 2003; McCarthy & Millard, 2003). As the NHS undergoes significant changes in terms of funding and organisation there will be specific changes that will impact on this group, therefore, providing feedback on potential services changes is important. For example, paediatric cardiac services are undergoing a public consultation (NHS, 2011), the outcome of which may have considerable implications for people with Down syndrome and their families: local centres may close; those commencing treatment may have further to travel; and more specialised teams may have greater expertise in working with people with care needs such as those with Down syndrome. However, unless the needs of this group are taken into account as part of the consultation, relevant improvements may not

happen. By incorporating views such as those elicited in the current research, changes to service provision may address current shortfalls and ensure future changes do not create further barriers to accessing services, where possible

This research has general implications for considering the role that family members and others play in consultation and therapeutic interventions with people with Down syndrome. While people should have the opportunity to represent themselves, there may also times where they are others who can collaborate or better represent their interests. This will be particularly important in the role of clinical psychologists in community learning disability teams as services move towards individual budgets and direct payments. Without knowledge of available choices, individuals will find it difficult to make choices. Independent advisors may be able to provide unbiased advice, but families and friends will be important advisors as people who know their family members best.

The research findings indicate that there are a variety of factors that may have an impact on satisfaction with cardiac care including familiarity with the cardiac team and a balance between consulting the individual and consulting their family members. Factors found, by wider research, to be central to successful therapeutic management of congenital heart disease include necessary medical knowledge and close contacts with medical teams (Kovacs, Silversides, Saidi, & Sears, 2006). One change proposed by a recent consultation document (NHS, 2011), is to improve antenatal diagnosis and support of congenital heart difficulties by local services. It is proposed that at diagnosis families:

..would have access to a clinical psychologist, nurse counsellor or specialist nurse. This is to ensure the necessary support and guidance is provided from the moment the child is diagnosed to enable parents to make informed decisions about care for their child (NHS, 2011, p. 56).

This recognition of the potential difficulty of the decision making process, the need for support for families, and role of the clinical psychologist in providing support, suggests that teams are ready to listen to families and afford them time and resources to make difficult decisions. Clinical psychologists are well placed to provide support in such circumstances and manage relationships between families,

diagnostic teams, and cardiac teams. However, to address the needs of adults, such as those consulted in the current study, this role will need to stretch beyond diagnosis, birth and decisions in early life. In addition, the clinical psychologist has been proposed to have important roles in multidisciplinary research and professional education within adult congenital heart disease services (Kovacs, Silversides, Saidi, & Sears, 2006). The breadth of clinical psychologists' training should ensure skills in working therapeutically with people with a learning disability, rather than needing to refer individuals to a community learning disability team, where psychologists will have less expertise in the care of cardiac health.

Limitations

Qualitative research in general has been criticised as being unscientific for a variety of reasons including small sample sizes, anecdotal qualities, and researcher bias, all of which (it is claimed), contribute to limited generalisability of research findings (Mays & Pope, 1995). Others, however, argue that the reduction of power imbalances between researchers and the researched encourages authenticity and honesty, which exposes power imbalances and provides a more, not less, accurate account of relationships (Karnieli-Miller, Strier, & Pessach, 2009). Quantitative research and some qualitative methods attempt to hide behind large numbers and large swathes of data in an attempt to prove they have uncovered a truth, however, this process can never be exhaustive. We can never include all possible relationships and permutations, therefore, the small details found in individual interactions may be as important as an amalgamation of the views of a much larger group where the subtleties of everyday life have been lost in the mix (Sacks, 1984). Ethnomethodology, for example, seeks to use normally occurring data to understand social action that is missed by more traditional statistical methods on the premise that "a detailed examination of even the tiniest fragments of the social order reveals important properties of the whole" (Rapley, in press).

Applying such a defence to the current research topic, to understand the experiences of people with Down syndrome and a CHC, we must first know what we are looking for, and the only way to ensure we do not miss important and subtle details is to adopt a broad and also detailed approach. While we aimed to achieve that in the current project, we do not know whether the experiences we heard about

are similar for other people with Down syndrome and a CHC. Consultation with the director of the DHG support group confirmed that information on people's experiences was needed in order to decide whether service improvements were required. There was anecdotal evidence on both a need for service improvements and examples of good practice, however, the DHG had neither the resources nor the expertise necessary to embark on such a research project and a skilled researcher was required to advance this knowledge further. The current method allowed the support group and participants to have their say without undue requirements being made of their time. The researcher will provide participants with a summary of findings and the support group will also receive input regarding results and how the group might use this information in the future. Such ongoing contact will help ensure that the write-up, any publications, and future projects keep the participants at the centre of this research as stakeholders and consultants.

Recent decades have seen a move towards the adoption of participatory methods in the realm of learning disability research (Chappell, 2000). The current project involved the participation of people with Down syndrome rather than focusing exclusively on the views of parents or carers. Participatory action researchers would, however, highlight the limitations presented by this type of approach. Participatory and emancipatory methods aim to allow people, not traditionally seen as researchers, to set the research agenda, identify areas important to them, and decide what questions should be addressed by research carried out in collaboration with skilled researchers (Baum, MacDougall, & Smith, 2006). In the current project, ethics procedures related to the clinical psychology training doctorate required that ownership of the project remain with the researcher. Further, the participatory research movement is in turn criticised for the challenges it creates for the realm of health and social care, given the potential that important areas are under researched. Gilbert (2004) outlines the developments made in making research projects and research practices more accessible and highlights that the research process, regardless of attempts to becoming participatory will always contain inherent power imbalances and be unable to include all individuals in any marginalised group (such as those with severe and profound learning disabilities). He suggests that 'as a commitment to an ethic of participation, researchers will need flexibility and patience' (Gilbert, 2004).

To demand equal participation from participants assumes that those affected will be interested in and motivated to develop and take part in research. Without the knowledge that service improvements are possible, or available, people with Down syndrome and a CHC may not be motivated become involved in research. This creates a vicious circle that illuminates the limits to all participatory research projects by their very existence, indicating that even in apparently empowering contexts groups can be disempowered and neglected as the research will be instigated by outsiders (Antaki, Finlay, & Walton, 2007). Therefore while participatory research methods have many positive aspects, for those with a learning disability, participatory methods may not always be inclusive.

Participatory research is closely aligned with the social model of disability, which claims that people are disabled by society and not their bodies, and disability is an experience rooted in the barriers people face in society (Office for Disability Issues, n d). This approach has been criticised, however, for its neglect of the reality of impairment for those with a disability. People may be disabled by society and by their bodies (Shakespeare & Watson, 2001), and impairment will be salient for many regardless of barrier removal: "We are not just disabled people, we are also people with impairments, and to pretend otherwise is to ignore a major part of our biographies" (Shakespeare & Watson, 2001, p.14). Impairments come in many shapes and sizes and affect people differently, in addition, individual preference will result in a variation in terms of interest in research. To assume participation in research is important to those with a disability prioritises goals of research over the reality of life with a disabling impairment, a reality that can render people unable, unwilling, or simply uninterested in participating. To propose, as emancipatory and participatory research do, that learning disability research should not take place without people with a learning disability is a dangerous and potentially debilitating view and risks that less, not more, will be learned about learning disability in the years to come. Participatory research, therefore, has positive and negative implications for research in this area. This project adopted the 'ethic' of participatory research (Gilbert, 2004), while also recognising that disabilities can be impairing and research can be intellectually demanding.

Attempts to provide accessible recruitment literature and make results accessible to participants highlight further limitations of this project. This research

required participants to have sufficient verbal skills to complete an interview, and, where any communication difficulties arose, mothers aided communication between researcher and participant. This design excludes those with more severe and profound learning disabilities and potentially those who did not have the support of parents and carers. We do not know what such participants may have said and, as with all research, the most vulnerable may have chosen not to take part (Bond-Sutton, Erlen, Glad, & Siminoff, 2003). Attempts have been made to measure the well-being and emotional responses of those with severe and profound learning disabilities (Vos, De Cock, Petry, Van Den Noortgate, & Maes, 2010), however, this input will be qualitatively different from the input that those with more developed verbal skills are able to contribute. This presents an ethical dilemma as we also know that staff tend to “seriously” over estimate client experiences and emotions, including empowerment, satisfaction, and sense of belonging (Rapley, Ridgway, & Beyer, 1998). Considerations of how to include feedback from all service users needs to consider how those with severe and profound learning disabilities could contribute meaningful feedback to NHS services, or whether agendas to pursue this is potentially a fruitless exercise that underestimates the reality of the impact of disability on the individual. The assumption that all are able to contribute to research fails to distinguish between social disability and biological impairment (Shakespeare & Watson, 2001).

The barriers encountered highlight the difficulties faced when addressing the gap in the evidence base and attempting to complete research with people with a learning disability rather than writing about them. In the current project a compromised between truly participatory research and research that could be clinically relevant to clinicians and people with Down syndrome. The research team employed a traditional method of researcher designed and lead interviews. Dilemmas encountered were held in mind in terms of recruitment, method, and outcomes of research. It also had an impact on the analysis of findings. Goodley and Rapley (2002), explored the development of the myth of “acquiescence bias” from Sigelman’s initial paper on the subject (Sigelman, 1981). They track its development from methodologically unsound beginnings, as it grew into a widely accepted concept, whereby it is assumed people with a learning disability will frequently say yes regardless of question content, and where inconsistent responses are viewed as

lack of comprehension, when in other groups this may be labelled disagreement (Goodley & Rapley, 2002). They explore how this concept can be created in interactions, in particular when interviewers come armed with schedules of questions. The interviewee could quickly come to the assumption that there are a range of possible right answers that they are expected to know and feel in an inferior position compared to the interviewer.

The findings of this research have highlighted that, when offered the opportunity, people with Down syndrome and a congenital heart condition are interested in and have the ability to share accounts of living with a cardiac condition. The themes identified and the relationships illustrated in accounts will have implications for other people with similar conditions, for teams developing their skills base and for families embarking on their own journey through the healthcare system.

Chapter 5 – Critical Reflections

The findings outlined above have implications for people with Down syndrome and their families in terms of how people both access and support medical decisions and decisions around health generally. They also have implications for the practice of healthcare professionals in relation to this group, and specifically for the practice of clinical psychologists and how they work therapeutically with people with Down syndrome and their families. The research has highlighted the important implications for including people with Down syndrome in active research. A review of the literature related to the inclusion of people with a learning disability and/or Down syndrome in research made direct suggestions for how research should be carried out with this group (e.g. Nind, 2008). Tips from previous research and advice given by parents prior to and during interviews noted that open questions would be more difficult for interviewees, as would thinking abstractly and the interviewer should be mindful of possible agreement with yes/no questions. All of these recommendations take the view that people with Down syndrome might find the process of interviewing confusing and that they would need to be primed for information. It suggests that researchers should disregard the Mental Capacity Act, which states that “a person must be assumed to have capacity unless it is established that s/he lacks capacity” (DCA, 2005, p.20).

These reflections, therefore, have their roots in the conception of the study and the assumptions that are made about people with Down syndrome. A thematic analysis of the interview data highlighted a fourth theme related to the process of taking part in research: “Don’t have a Scooby” – making consent meaningful. The inclusion of the theme here aims to illustrate the additional challenges that face people with Down syndrome when they are making themselves heard.

“Don’t have a Scooby” – making consent meaningful

There were times where the answer “don’t know” was given where further questioning revealed interviewees had more knowledge than they were initially able, or willing to verbalise. In addition, there were points where *don’t know* responses were articulated through the use of other phrases such as “I don’t have a Scooby”, used by two participants:

I: So maybe [the dietician] tells you you need to eat smaller dinners does she?

Sam: Yeah

Sam's mother: Yes

I: And why does she say that?

Sam: Cause of Bella [family pet dog]

I: Why does she say that Sam, why does she to have smaller dinners?

Sam: I don't have a Scooby

This corresponds to the rhyming slang phrase *I don't have a Scooby Doo* meaning "I don't have a clue" and refers to Scooby Doo, a children's TV cartoon character. At other times interviewees made suggestions that the interviewer should ask mothers for further information, which also indicated that they would rather change to different topics of conversation:

I: Who is Louise? Is she your sister?

Adam: Mm hmm

I: And what age is she?

Adam: Eh, don't know

I: Is she older or younger than you?

Adam: I don't know, you have to ask Mum

I: Have to ask Mum, well maybe we'll ask Mum later

Adam: Mmm hmm

Both strategies resulted in the interviewer not pursuing the avenue of questioning further and are interpreted as examples of interviewees exercising choice with regard to involvement.

There were times when interviewees struggled to understand concepts in particular when trying to elicit opinions about views held by others. The interviewer had to balance the valid pursuit of questioning with the possibility that interviewees may feel coerced or pressured into answering:

I: And so it sounds like Mum knows all about your health, what about when you are not feeling well, what do you do?

Adam: Em I don't know

I: Do you tell Mum or tell you sister (mm hmm) What do you do

Adam: I don't know

I: Do you tell Mum?

Adam: Yeah

I: What do you tell Mum?

Adam: Em

I: Might you say I'm not feeling well

Adam: Little bit

I: A little bit, and what will Mum do?

Adam: Em.. hmm

I: Does she call the GP or do something else

Adam: I don't know.

Interviewees verbalised anxieties around taking part in research and some reported having discussed their anxieties with others, demonstrating the careful consideration that had gone into interview preparation, particularly for Katherine and Jane. Checking in at the end of interviews indicated that both women had found the interview less anxiety provoking than feared:

I: Good and so if you are worried about anything do you tell Jane about it?

Katherine: Yeah

I: What kind of things do you talk to her about?

Katherine: Em well being nervous about this

(...)

I: What was it like answering my questions?

Katherine: Quite nervous

I: Quite nervous? And how to you feel now?

Katherine: Okay

I: you feel okay now, so it wasn't too scary....it wasn't too bad after all?

Katherine: No

They stated that having the interview together would have been 'better', but also agreed that had they been interviewed together they would have been distracted. Such comments highlight the emotionality of research participation. One-

to-one, semi-structured, one-off interviews can be nerve wracking and a source of distress despite eagerness and willingness to take part in research (Dickson-Swift et al., 2007). Alternative methods could go some way to allaying these fears, such as longer, or repeated sessions, however, ethics procedures demanded by the university limited the adjustments to research methods that could be made. Despite points of confusion and avoidance of some questions, interviewees did not choose to end interviews. Interviewees at times took charge of interviews by rebuffing mothers when they interrupted:

Tom: Yeah I know

Tom's mother: What happens? Are you okay? Do you get pain?

Tom: Mum me not you.

And the use of humour suggested an enjoyment of being interviewed:

I: And what happens if the readings aren't between those? .. Does that ever happen?

Sam's mother: Eh yes, what do we do

I: What do you do then?

Sam's mother: When its four what do we do?

Sam: In the bin [exclaimed]

These reflections on the interviewing process are presented in order to give insight to the experience of being interviewed and some of the barriers and bonuses to including people with Down syndrome in research rather than carrying out research about them. Participants were happy to talk candidly about their experiences and it feels important to me as a researcher that views such as these are utilised in the design of healthcare services, not just as a commentary on past experiences. Previous service evaluation of cardiac services has failed to consider the needs of those with Down syndrome or a learning disability, for example the report on the Bristol Inquiry into children's heart surgery (Kennedy, 2001), has no explicit acknowledgement of the people with Down syndrome and their families who were affected by the scandal ("Bereaved parents 'treated shamefully'," n.d). Subsequent literature on consent procedures has not been made accessible for people with literacy needs and there are no official resources dedicated to people

with Down syndrome who have cardiac needs. The failure to involve people with Down syndrome in service evaluation misses the potentially valuable contributions demonstrated by the current research. It will also result in a lack of healthcare team expertise about working with people with Down syndrome, once again requiring parents to be a source of expertise and potentially relying on family members to make medical decisions, when such decisions could be supported instead by an Independent Mental Capacity Advisor (IMCA) (DCA, 2005), in conjunction with the medical team.

These reflections and the links that have been made to existing literature, are designed to provide a framework for understanding how these five people with Down syndrome and a CHC constructed their experiences. The process of analysis and write-up has been evaluated in line with the thorough checklist of criteria proposed by Braun and Clarke (2006, Appendix I), to evaluate the process and products of a good thematic analysis. While thematic analysis can pose a disadvantage in that the breadth of the approach means many things can be said about the data, and focusing on specific aspects can, therefore, be difficult, there are many advantages to the method, particularly for clinical psychologists. As a relatively easy and quick method to learn, thematic analysis is accessible to participatory research, with participants as collaborators, an application not achieved in the current research. It is flexible to multiple research topics, and data sets, and allows researchers to report on similarities and differences across the data set, which has been a key characteristic of the current research. The method allows the voice of people with Down syndrome to be heard and will help inform future service development in an area, which has received little attention thus far. "Public health policy and health promotion strategies need to include the particular challenges faced by persons with disabilities if the latter are to fully share in achieving health gain" (Noonan-Walsh & McConkey, 2009, p.33).

There were other areas, which were not as sensitive to the difficulties people with Down syndrome might experience when accessing research. Time and relationship building have been cited as two key considerations in the development of truly participatory research (Nind, 2008), and are necessary to avoid "parasitic" practices long condemned in disability research (Stone & Priestley, 1996). The ethics process was inflexible in terms of the order in which consultation and planning for

project design could proceed. While the clinical doctorate programme provides a significant time period for the completion of research, following literature reviewing and consultation with various accomplished participatory researchers in the Greater London area, it became apparent that this time scale was not suitable for participatory research and it would be unfair to inflict the limited flexibility of what was on offer on potential fellow researchers. “Probably the most controversial [concern for the social researcher] relates to the contention that researchers must be accountable to disabled people and their organisations. However, to be accountable to the entire disabled population would be impossible” (Barnes, 2003, p.5). As an employee of the NHS I could not transfer ownership and direction of the research to participants, nor would I aim to represent the views of all people with Down syndrome and a CHC. The project that resulted held these difficulties in mind to ensure the research maintained, at the very least, a balance between the demands of the clinical doctorate, demands of research procedures, and the needs of the participants.

By holding the decisions made in mind, risk was minimised. However, the potential for harm due to research participation can never be removed completely. Measures can be taken to minimise risk and therefore protect the rights of people, in particular vulnerable groups to take part in research (Bond-Sutton et al., 2003). Good communication, clarity regarding conflicts of interest, meaningful consent procedures, and simplification of research procedures could all help with this. Responsibility should be shared by researchers, funders, ethics committees, and participants alike and the University could do more to learn about vulnerable groups in order to better understand the implications of granting or denying ethical approval and encouraging research in the area. Additionally, as researchers, we should consider what it would mean for people with a learning disability to be empowered as researchers (Duckett & Fryer, 1998). Given freedom and a research agenda, would they choose to embark on any research at all, do they see themselves as worthy of being the subject of research (Brown et al., 2010), or are researchers by their very instigation of research placing people in a disempowered position. The plan is to take this research further by continuing to consult with research participants and allow their views to inform future research directions. Additionally the writer will

integrate research into clinical psychology practice to help ensure research agendas are pursued where knowledge and literature are lacking or untested.

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Appendix A
Information Sheet

Dear



My name is Deirdre



- I am writing about what happens when people have heart problems.
- Some people with Down syndrome have heart problems.
- They go to the GP
- Sometimes they go to hospital



-
- I want to talk to people about what it is like to have Down syndrome and heart problems
 - Do you want to help me do this?
 - I will meet with you and we will talk
 - I will write things down, but I will not use your name.
 - You will not get in trouble if you do not want to help me.

✓ You can say YES or No

X It's up to you

To find out more:



- You can talk to (named service contact)
- Call me on 0208 533 9616
- Email me on d.reilly@nhs.net

If you have questions worries about the research you can contact my supervisor at the university –

Mark Rapley 0208 223 4174

✂.....

If you want to talk to me call me on 0208 533 9616

OR complete the form and post it to me:



Deirdre Reilly
School of Psychology
UEL Stratford
London
E14 4LZ

Your Name and Address

.....
.....
.....
..... ①

Appendix B

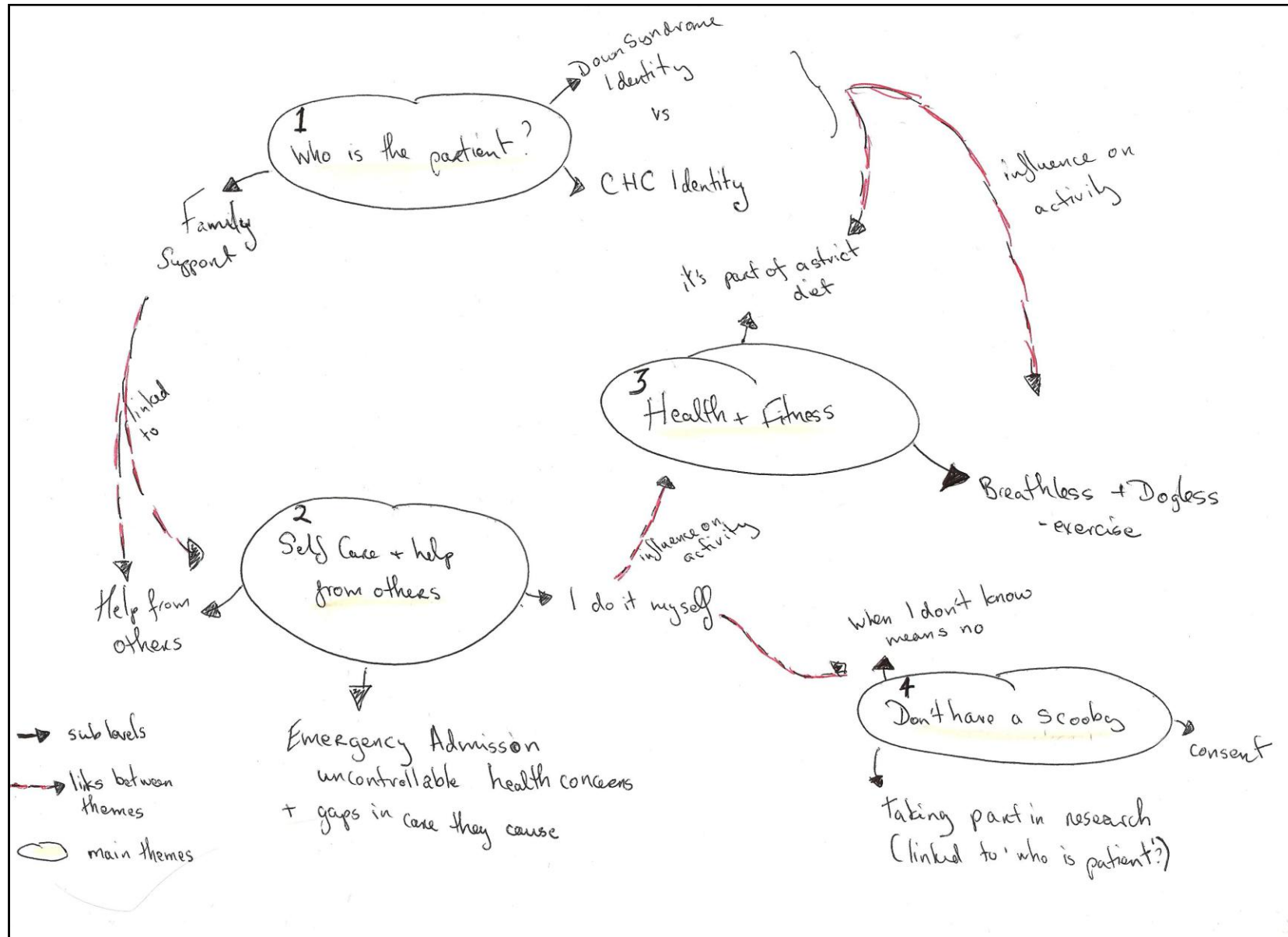
Sample from transcribed Interview

		Initial thoughts + notes	<u>Codes</u>	<u>Themes</u>
1	DR: and are you able to do those things with your heart?			
2	█: umm			Managing own health
3	Mum: do you have to go fast or have to go slow?			
4	█: <u>Go slow yeah</u>	limits what to do when poorly	limits	
5	DR: you have to go slow? And why do you have to go slow?			
6	█: umm			
7	Mum: what happens if you do too much dancing?			
8	█: not sure.			
9	Mum: what happens if you do <u>too</u> much dancing?	over exertion		
10	█: yeah I know			Involvement interested
11	Mum: what happens? Are you okay? Do you get pain?			
12	█: Mum me not you.	wanting to tell story - Mum taking over	process note.	
13	Mum: so what happens?		body control	
14	█: I don't know	understanding	understand question / concept	
15	Mum: you don't know			
16	DR: so does it get sore, does your heart get really fast? DO you get pain?		limits due to heart	Managing health
17	█: <u>I get pain</u>	pain		limitations to activities
18	DR: You get pain if you do too much dancing. And what about too much football?			

Extract 62

Appendix C

Example of Theme Spider Diagram



Appendix D

Thematic Analysis Candidate Themes

1. Down syndrome and lifestyle
 - a. Knowledge and meaning
 - b. Identity and friendships
 - c. Education
 - d. Family support
 - e. Lifestyle/ future
2. Down syndrome as a barrier to independence
 - a. Good and bad things about DS
 - b. Special treatment
3. Health
 - a. Birth history/ history of illness
 - b. General health/GP
 - c. Hospital health care/ relationship with team
 - d. Self Management vs Mum in charge
 - e. Co-morbidity
 - f. Healthy eating and fitness
4. Heart
 - a. Knowledge: surgery, check-ups, current treatment
 - b. Help seeking
 - c. Hospital/ Emergency admission/ relationship with team
 - d. Symptom management
5. Ask Mum
 - a. Parental responsibility for health
 - b. Mum steering interview
 - c. Send to Mum for further info /asking mum to back off
6. Don't have a Scooby
 - a. Knowledge gaps

Miscellaneous /Process: Tricky concepts

Changing topics

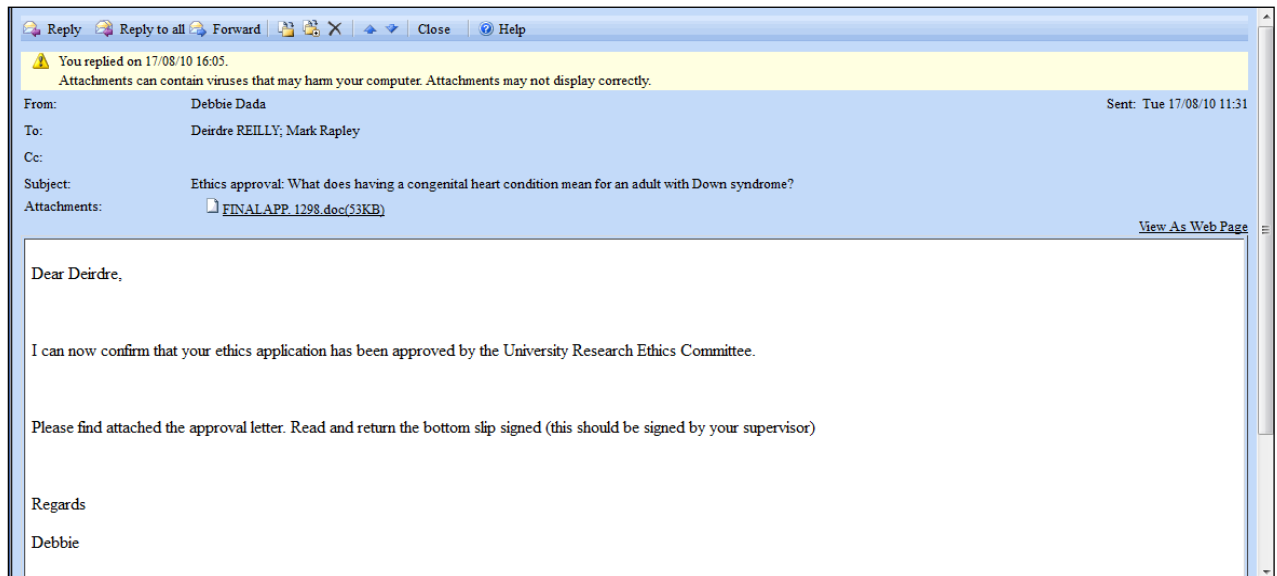
Confusion

Humour

Anxious of performance

Appendix E

Confirmation of ethical approval





Professor M Rapley
School of Psychology
Stratford

ETH/12/98

06 May 2011

Dear Professor Rapley,

Application to the Research Ethics Committee: What does having a congenital heart condition mean for an adult with Down syndrome? (D Reilly)

I advise that Members of the Research Ethics Committee have now approved the above application on the terms previously advised to you. The Research Ethics Committee should be informed of any significant changes that take place after approval has been given. Examples of such changes include any change to the scope, methodology or composition of investigative team. These examples are not exclusive and the person responsible for the programme must exercise proper judgement in determining what should be brought to the attention of the Committee.

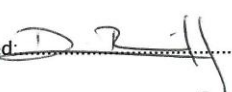
In accepting the terms previously advised to you I would be grateful if you could return the declaration form below, duly signed and dated, confirming that you will inform the committee of any changes to your approved programme.

Yours sincerely

Debbie Dada
Admissions and Ethics Officer
Direct Line: 0208 223 2976
Email: d.dada@uel.ac.uk

Research Ethics Committee: ETH/12/98

I hereby agree to inform the Research Ethics Committee of any changes to be made to the above approved programme and any adverse incidents that arise during the conduct of the programme.

Signed:  Date: 17/8/2010

Please Print Name:

DEIRDRE REILLY

Appendix F
Consent Form



Life with a heart condition

Consent form

NAME:



Tick if agree

I want to do an interview


☐

I know what we will be talking about


☐

I know I can end the interview at any time


☐

I know that if I have any questions I can speak to Deirdre Reilly



Tel - 07841430221

☐

Signature:.....

Date:.....

Researcher signature:.....

Date:.....

Appendix G

Interview Schedule

Recollection of initial meeting	<p>Do you remember what we talked about during the last time we met?</p> <p>Do you know why I want to talk to you today?</p>
General	<p>Questions about you and your family</p> <p>Tell me what you like doing?</p>
Down syndrome and intellectual disability	<p>What is Down syndrome?</p> <p>Do you know anybody who has Down syndrome?</p> <p>What do you think having Down syndrome means? What is DS like?</p>
Heart problems and health	<p>When we last met, we talked about people having health problems. Do you remember what we said about that? Can you tell me about your health/heart problem?</p> <p>What is it like having a heart condition. What does it mean for you?</p> <p>Do you have a hospital passport?</p>
Primary health care and Down syndrome	<p>When we last met, I told you I would like to talk to you about seeing your doctor ? Do you remember what we talked about?</p> <p>Have you seen your doctor to talk about your heart problem?</p> <p>Do you have a health action plan or hospital passport?</p> <p>What is it like going to the doctor? Is there anything you would like to change about going to the doctor?</p> <p>Tell me about a time you went to the doctor?</p>
Secondary health care and Down syndrome	<p>Have you been in hospital? What happened in hospital?</p> <p>Tell me about a time you were in hospital?</p> <p>What did you like/dislike about hospital?</p> <p>What could make hospital better/worse?</p>
Other questions	<p>We talked about what doctors know about Down syndrome, do you remember what I said?</p> <p>What do you think doctors know about Down syndrome?</p> <p>Is there anything you think doctors/nurses need to know about people with Down syndrome</p> <p>Is there anything you would like to tell your doctors about you?</p> <p>Is there anyone else who helps you go to the doctor or tells you about health issues?</p> <p>Where else to you get information from?</p>

Appendix H
Demographic Information

Demographic information

(To filled in by researcher during interview)

Participant number _____.

Date of interview _____.

Age: _____

Gender:

Male ☐ Female ☐

Physical Health (report indicate whether self or other)

Excellent ☐ Very good ☐ Good ☐ Fair ☐ Poor ☐

Heart condition and brief history

Ethnicity: (tick box)

English	Welsh	Scottish
African	Afro-Caribbean	Bangladeshi
Chinese	Indian	Irish
Other European	Pakistani	Other _____ <input type="checkbox"/> .

Present employment status?

Employed full-time ☐ In full-time education ☐ Unemployed ☐

Employed part-time ☐ Voluntary work ☐ Retired ☐

Details:-

Current residential/marital status:

With one parent Y/N male/female

With both parents Y/N

Residential/educational placement Y/N

Supported accommodation Y/N

Independent Y/N

(With partner Y/N, With Children Y/N)

Family

If living with family who lives at home?

In total how many: Brothers _____. Sisters_____.

Place in family_____.

Do any of siblings have a disability? Y/N

Details_____.

Interests

Future plans

Appendix I
Checklist of Criteria for Good Thematic Analysis

Table 2 A 15-point checklist of criteria for good thematic analysis		
Process	No.	Criteria
Transcription	1	The data have been transcribed to an appropriate level of detail, and the transcripts have been checked against the tapes for 'accuracy'.
Coding	2	Each data item has been given equal attention in the coding process.
	3	Themes have not been generated from a few vivid examples (an anecdotal approach), but instead the coding process has been thorough, inclusive and comprehensive.
	4	All relevant extracts for all each theme have been collated.
	5	Themes have been checked against each other and back to the original data set.
Analysis	6	Themes are internally coherent, consistent, and distinctive.
	7	Data have been analysed – interpreted, made sense of – rather than just paraphrased or described.
	8	Analysis and data match each other – the extracts illustrate the analytic claims.
	9	Analysis tells a convincing and well-organized story about the data and topic.
Overall	10	A good balance between analytic narrative and illustrative extracts is provided.
	11	Enough time has been allocated to complete all phases of the analysis adequately, without rushing a phase or giving it a once-over-lightly.
Written report	12	The assumptions about, and specific approach to, thematic analysis are clearly explicated.
	13	There is a good fit between what you claim you do, and what you show you have done – ie, described method and reported analysis are consistent.
	14	The language and concepts used in the report are consistent with the epistemological position of the analysis.
	15	The researcher is positioned as <i>active</i> in the research process; themes do not just 'emerge'.

(Braun & Clarke, 2006, p.96)